Ankylosing Spondylitis

CLINICAL
CONSIDERATIONS ROENTGENOLOGY
PATHOLOGIC ANATOMY TREATMENT

By

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Preface to the American Edition

This book is devoted essentially to a clinical description of the condition, Ankylosing Spondylitis, in all the phases of its course Even the title indicates that we have considered this disease as distinct from rheumatoid arthritis, an opinion which is not shared by the majority of American rheumatologists Some biologic studies seem to furnish evidence in favor of such a distinction

From the French edition of 1951 we have made only changes in presentation We have also added to the bibliography papers that have come to our attention since 1950, the original bibliography having ended at that time

We would like to emphasize one point in the presentation which we adopted for the first edition The frequency and the time of appearance of the symptoms have been based on a study of 200 cases. Therefore, some of the cases were seen at the beginning of the disease, while others were not seen until the disease had reached an advanced stage we have used this heterogeneous series of cases, because these cases reflected the diagnostic problems that were brought to us In order to facilitate the reading of the book, we have presented our findings in the form of percentages, but these figures are not to be regarded as statistics A series of cases of Ankylosing Spondylitis of more than 20 years duration would have permitted us to give more precise data on the frequency of different manifestations as they appear during the course of the disease, but the frequency mentioned in the book is based on only 40 cases

We wish to express our gratitude to Doctor A U Desjardins, who spontaneously undertook this translation and who has carried out this task with a conscientiousness and faithfulness which we cannot praise sufficiently

J F
F J
J R

Translator's Preface

Translation of a book from one language into another is not as easy as it may appear to readers who know only one language. When the book is a literary work, and especially one of high quality, the task is all the more difficult, because the translator must not only render accurately the meaning and ideas of the author, but he should also be able to transmit to the readers the thought and lilt of the poetry or the vigor and fluency of the prose — in other words, the readers should obtain the "literary feel" of the original text and this is quite a task for anyone to undertake.

As for a book on a technical subject, the translator's first object should be to make certain that he understands what the author has tried to say or to explain; it is more important to make out the intended meaning than to pay too much attention to the actual words that may have been employed, because sometimes the words that have been used do not accurately convey the meaning. When the meaning is thoroughly understood, the translator's only remaining task is to convey this meaning to the readers in as fluent, clear and readable language as he can find. Usually this is not difficult because, when the meaning is clear, the proper words to express it should come almost unbidden. Boileau, the poet, once expressed this thought perfectly:

Ce que l'on conçoit bien
S'énonce clairement
Et les mots pour le dire
Arrivent aisément

When I undertook to translate this book, which had been written by my friends, Doctor Jacques Forestier, Doctor Jacqueline, and Doctor Rotes-Querol, I realized at once that my efforts should be directed, not toward trying to make a literary production out of a purely technical, and in this case medical, text, but to render the

thoughts, ideas, and explanations of the authors as clearly and as fluently as the English language made this possible. How nearly I may have succeeded in accomplishing this remains to be seen I am painfully aware that, however good may be one's intentions and however hard one may labor, errors have a pernicious way of creeping into any human effort I am not vain enough to deceive myself into thinking that I have escaped this danger altogether, but I hope that any errors that may have crept in may be few and not too serious, because once the book is "out," the verdict must remain with the readers, who often are not chary about expressing their disappointment or disapproval And this is as it should be After all, anyone who undertakes to translate a book should know enough about the subject and about books in general to steer clear of serious blunders I bow my head in humility and in hope

Walpole, Maine ARTHUR U DESJARDINS, M D

Preface

Ankylosing Spondylitis is a disease which is frequently observed in the practice of rheumatology Our knowledge of it as a clinical entity, however, is of relatively recent date, because hardly more than half a century has elapsed since the group of spinal stiffenings and ankylosis began to be dissociated But since then many descriptions of the symptoms and principal features of the disease have been published

We have personally gathered more than 400 cases of this condition This observed material is homogeneous, because it has been *gathered into groups* by one of us, moreover, it does not consist of instantaneous impressions" obtained at a single examination indeed, detailed histories of these cases have been obtained and, what is still more important we have been able to follow a large proportion of the patients for many years, thus we have been able to observe the evolution of these cases, whether they had received treatment or not, and many have been followed from the onset of the disease All these data, in connection with which Doctor Certonciny has rendered us invaluable assistance, has enabled us to acquire a better knowledge of the condition and to formulate a personal conception of it. In this book we will set forth our own opinions which, as we have just mentioned, are based on personally observed facts, and compare them with those of other authors

Since our present knowledge does not permit us to draw any firm conclusions, the problems of etiology and pathogenesis will be simply set forth.

We wish to extend our thanks to Doctors P Robert and P Faidherbe, whose roentgenograms comprise nearly all of the roentgenographic illustrations in this volume to Professor Rutishauser and his pupils, Doctors Riotton and Wettstein, who have placed at our disposal records of their pathologic studies to Doctor J E W Brocher who collaborated with one of us in the preparation of a

monograph on Ankylosing Spondylitis and to Professor Franceschetti who helped us with the chapter relating to the ocular symptoms.

We also thank the publishers, Masson and Company, for having published this book and for having permitted us to illustrate it freely

J F
F J
J R

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Ankylosing Spondylitis

Historical Background

Although certain forms of rheumatism and gout have been known since antiquity, this has not been true of Ankylosing Spondylitis, which was first studied on anatomic material at the end of the XVIIth century

1691—Bernard Connor (43), an Irishman, wrote, at the Faculty of Rheims (France), a thesis entitled 'Sur la continuité de plusieurs os, à l occasion d un tronc de squelette humain où les vertèbres, les côtes, l os sacrum et les os des iles, qui naturellement sont distincts et séparés, ne font qu un seul os continu et inséparable" From this anatomic study he concluded that this person must have been incapable of motion, that it must have been impossible for him to bend to stretch, to go to bed or to rise from it, or to turn on his side and that his respiration must have been very limited In this description we find the features which, even today, are considered the most important symptoms of Ankylosing Spondylitis

1874—Hilton Fagge (57) gave, before the Surgical Society of London, the report of a necropsy on a subject whose body presented a complete ankylosis of the spine, he drew special attention to an intense osteoporosis of the vertebrae, the bodies of which could be cut with a knife, in contrast to the hardness of the ligamentary ossifications Still it is impossible to be certain that the disease in this case was true Ankylosing Spondylitis

1884—A Strümpell, in his *Manual of Internal Medicine* (208), described for the first time and in the following terms the disease with which we are now concerned ' Comme une affection remarquable et, semble t il même, particulière, on peut signaler encore ici, en passant, cette forme morbide dans laquelle survient, tout progressivement et *sans douleur*, une ankylose complète de tout le rachis et des deux articulations coxo-fémorales, de telle sorte que la tête, le tronc et la cuisse sont intimement soudés ensemble et tout à fait rigides, tandis que les autres articulations conservent

leur mobilité normale. Il va de soi que, par suite de cette soudure des modifications toutes particulières de l'attitude et de la démarche prennent naissance. Nous-mêmes avons vu deux cas tout à fait semblables de cette singulière affection "[1]

1893—W Bechterew (10) described a chronic condition of the spine which was characterized by diffuse and considerable stiffening, by kyphosis especially in the upper dorsal region, and by neurologic manifestations incidental to compression of nerve roots associated with muscular atrophy and sensory disturbances. He reported three cases in which it is difficult to be certain whether or not Ankylosing Spondylitis was present, and in two of them he found *hereditary and traumatic factors* By bringing together syndromes of different origin and character Bechterew does not appear to have contributed anything that might have helped to distinguish Ankylosing Spondylitis from the general group of spinal diseases Nevertheless, in the English and German literature, his name has remained as a sort of landmark.

1897—In the same issue of the *Deutsche Zeitschrift für Nervenheilkunde*, Bechterew (11) and Strümpell (209), one after the other, took up the disease anew Bechterew repeated his previous description, and Strümpell reported a fresh case Besides the features which had previously been described, Strümpell noted the existence of *lumbar* and *sacral* pain But because of the absence of kyphosis and of radiating pain in his three cases, the latter author thought they were examples of a different condition from that which Bechterew had described

1897—Pierre Marie and Ch Astié (142) presented the case of an aged patient with a high dorsal kyphosis which had become distinctly aggravated after a fall The father and sister of this patient were more or less round shouldered Marie and Astié, perhaps a little too blithely, classified this case with the two cases

[1] As a remarkable and, it would seem, peculiar condition, we might now draw attention, in passing to a form of morbidity in which there supervenes, progressively and *without pain*, a complete ankylosis of the entire spine and of both ilio-femoral joints, in such a manner that the head, the trunk and the thighs become fused to one another and completely rigid, while the other joints preserve their normal mobility It goes without saying that, as a result of this fusion, very peculiar alterations in posture and gait develop. We ourselves have observed two absolutely similar cases of this singular condition.

which had been reported by Bechterew and proposed as a designation the term "*Heredo-traumatic kyphosis*"

A necropsy on this case, which had been performed later by Léri (136), had revealed ossification of the fibrous sheath which surrounds the spine—an ossification which is identical with that which will be described later under the name of "senile ankylosing hyperostosis of the vertebrae," it also disclosed one wedge shaped vertebra, which was the result of a fracture and which must have occurred at the time of the fall

Therefore, heredo-traumatic kyphosis" does not exist as an entity, but from the time that Pierre Marie and Astié reported this case, and especially in the French literature, the name of Bechterew has been attached to an ill-defined type of traumatic kyphosis that did not correspond to the cases described by Bechterew and that has since been copied uncritically in medical treatises

1898—Pierre Marie (143) reported two new cases of Ankylosing Spondylitis with a characteristic and very complete clinical picture He classified them with those of Strümpell and proposed the term of *Spondylose Rhizomélique*

1901—H Forestier (60), while describing more precisely the symptoms of Ankylosing Spondylitis, drew attention to the radicular involvement at all levels of the spinal column, and also to the importance of involvement of the inter apophyseal articulations

1903—Siven (199) was the first to study histologically a case of Ankylosing Spondylitis in a man aged 23 years. He described the alterations found in the interapophyseal and costovertebral articulations, and he concluded that these had an inflammatory character He also described the ossification of the intervertebral discs related to the same process

1904–1907—Fraenkel (82, 83) regarded the ankylosis of the interapophyseal articulations as characteristic of the condition he also studied the ankylosis affecting the costovertebral articulations as well as that of the sacroiliac joints He advocated the name of Spondylarthritis ankylopoietica "

1906—Pierre Marie and Léri (135) carried out a general clinical study of the disease and added six personal cases and one necropsy They described the ossifications of the ligamentary system, the ankylosis of the interapophyseal articulations and sacroiliac fusion,

although they did not ascribe to these two last alterations any particular value. Thus, since then, the anatomo-clinical picture of the disease has been well established They regarded the disease as an osteopathy

1918—Geilinger (92) emphasized the frequency of onset of the condition by involvement of peripheral joints (22% of the cases had had only a peripheral onset, and 52% had had at the same time a peripheral and a spinal onset)

1930—Kienböck called attention to the *bilateral sacroiliac synostosis* and thought of the *disease as a synostosing, articular inflammation*

1931—Krebs (122) noted that the *roentgenologic changes in the sacroiliac joints occur early and are characteristic* in 1934 Krebs and Vontz (123) *emphasized the same point and its diagnostic importance*

1931—Buckley (30, 31) published an excellent general study of the question, and in this study he rightly distinguished Ankylosing Spondylitis from other spinal disorders and emphasized osteoporosis of the vertebral bodies as an early diagnostic sign

1933—Güntz (98) gave a macroscopic and microscopic description of the anatomic and pathologic changes, and he concluded that the disease consists of an inflammation of the interapophyseal joints of the spinal column, ending in bony ankylosis, according to him, ossification of the ligaments would be a secondary manifestation

1934—J Forestier and P Robert (63) gave a more precise description of the roentgenographic appearance of the ligamental ossifications, and they advocated the term "syndesmophytes" which Sicard had suggested to one of them and which is to be used in opposition to that of osteophytes' which are characteristic of *degenerative disease of the spine*

1936—Gilbert Scott (191) in England, and M P Weill and Oumansky (215) and especially J Forestier (64) in France, divulged and emphasized the precocity of roentgenographically apparent involvement of the sacroiliac joints and of the diagnostic value of these findings which had already been pointed out by Krebs

1936—Campbell Golding (35), in a study of the initial clinical symptoms of the disease, emphasized their onset by involvement of peripheral joints

1939—J Forestier and J Metzger (70) studied roentgenograph

ically the evolution of the sacroiliac changes and described three successive stages

SYNONYMS

Among the numerous terms which have been proposed we will cite the following

1 Bechterew's Disease
2 Strümpell Marie Bechterew Disease
3 Spondylose rhizomélique (Pierre Marie 1898)
4 Spondylarthritis ankylopoietica (Fraenkel, 1904)
5 Spondylitis ossificans ligamentosa (Knaggs, 1924)
6 Fibrositis ankylopoietica dorsi (Krebs, 1931)
7 Syndesmite ossifiante (Simmonds, 1931)
8 Ankylosing Spondylitis (Buckley, 1935)
9 Spondylarthrite ankylosante (J Forestier, 1936)
10 Spondylitis adolescens (G Scott, 1936)
11 Rheumatoid Spondylitis (American Rheumatism Association, 1941)
12 Rheumatoid Arthritis of the Spine (a term often used in American literature)

As the most descriptive term we have decided to adopt the one proposed by Fraenkel *Spondylarthritis ankylopoietica*, or its French variant *Spondylarthrite ankylosante* (Ankylosing Spondylitis)

Incidence of the Disease

It is difficult to give even an approximate figure of the *morbidity of this disease for the human species* One cannot take as a basis the number of diagnosed cases which certainly is far below the number of existing cases.

Schmorl and Junghanns (189) found the vertebral columns of six or eight individuals affected with changes of Ankylosing Spondylitis out of 10,000 bodies which they had examined at necropsy but more than half of these spines had been examined only superficially by simple palpation at the time of necropsy It must be evident that, with this kind of examination, many cases may have been missed

H F West (218) studied the frequency of the disease in the population of Bristol (England), where he found that one inhabitant out of 2,000 was a victim of this form of Spondylitis

> In Bristol rheumatic diseases are centralized in the Royal Hospital, and it may be admitted that nearly all the cases that were diagnosed during the last five years had passed through this hospital Since only about 2% of the patients whom we have observed had had their first symptoms after the age of 40 years, it is possible, by means of a simple histogram, to make a rough estimate of the number of cases of Ankylosing Spondylitis in Bristol Considering the fact that the average age of death in patients afflicted with Ankylosing Spondylitis is 60 years, it is possible to say that the total number of known and suspected cases is 144 Since about 70% of the population of 420 000 inhabitants are insured the number of persons afflicted with this disease would be one in 2,000 of the insured population. This figure must be taken as less than the true incidence.

Although this estimate of incidence, as given by West, is only approximate, it seems to us to deserve some consideration

It is also difficult to specify the *incidence of Ankylosing Spondylitis in comparison with that of other rheumatic diseases* Boland and Present (19) found that 18% of the patients afflicted with chronic lumbar pain, who had been admitted to a general hospital of the American Army, had Ankylosing Spondylitis. Among the 1,000 most recent patients complaining of "rheumatism" in the broad sense of this word, who *composed the "spa clientele* of one of us in Aix les-Bains, the number of cases of Ankylosing Spondylitis was 25, while 69 cases of rheumatoid arthritis were observed From this observation it would appear that Ankylosing Spondylitis is only three times less frequent than rheumatoid arthritis. Without any doubt the generally accepted notion of the comparative rarity of this disease is related to the fact that a high proportion of cases escape statistical investigation because, in the absence of obvious spinal signs and because of the predominance of peripheral or radiating manifestations, the patients' symptoms are ascribed to other diseases, rheumatic or otherwise

Thus Ankylosing Spondylitis must be regarded as a relatively frequent disease and, since it persists throughout life without shortening it much, we have the explanation of the large number of cases that accumulate in our hospital services.

The disease affects all races and appears to be widespread in all countries. In Europe important statistics, including many hundreds of cases, have been reported Germany (Ehrlich 753 cases in 1930, in a health insurance company (55)), England (Buckley (32), Scott (192)), France (Forestier, (71)), Spain (Arcusa and Sans (1)), Sweden (Dekkers), Denmark (Overgaard (153)), Italy In North America the largest series was reported by Polley and Slocumb (158), and Ankylosing Spondylitis has been observed in all the other continents.

At the present time the commonly accepted *proportion, according to sex*, is 10 men to one woman (Polley and Slocumb (158)) In our statistics gathered from 200 cases (78) we have found that 82% of the patients were males, and 18% were females This proportion of 18% of female cases may be reduced to 14% or 16%, because our practice in Aix les-Bains is predominantly feminine nevertheless, it is the highest incidence of which we have any knowledge In women Ankylosing Spondylitis often goes unrecognized,

may have supervened long before the onset of the disease should be set at a still earlier date, in order to come closer to reality, therefore, one should displace toward the left the apex of the curve here presented

A fresh and more intensive study of cases with a late onset has led us to eliminate the last portion of the curve (from 50 years on) which had been prepared in 1949, because it doubtless corresponds to cases with a latent evolution and late outbreak like those which M P Weil and Sichere (217) have emphasized

Ankylosing Spondylitis in Animals Extensive spinal ankyloses in several species of animals have been described in the horse, the cow, the cat (the iron cat) (26), the dog but it is not certain that these ankyloses are identical with Ankylosing Spondylitis as it is observed in man

Until now, unfortunately, we have not been able to correlate our investigations with those of verterinary surgeons, and we regret not to be able to give even a few glimpses of the existence or non existence of ankylosing spondylitis in animals

General Considerations

Ankylosing Spondylitis is an inflammatory, rheumatic disease with a chronic evolution and an ankylosing tendency, which involves the entire locomotor apparatus, but particularly the articulations of the spine, those of the shoulders, and those of the lower extremities It may also affect the connective tissue of the eye.

Most frequently the disease *makes its' appearance* in men and during the adolescent period

From the clinical point of view we will divide our description into three periods which are arbitrary but which, for both practical and didactic reasons, seem useful to us

STAGE OF ONSET

This is the phase during which a definite and permanent limitation of mobility of the spine is not present By a definite limitation we understand a limitation which involves the loss of about a third of the normal mobility of one of the spinal segments, and by permanent limitation, the fact that it is present, not only during painful episodes, but also during the remissions.

Usually this phase of onset is *insidious* and intermittent, the symptoms vary and are not characteristic for this reason it is often difficult to determine which were the first symptoms of the disease According to our clinical observations, they have been as follows

Pain around the spine lumbar or sacroiliac, inflammatory in type, and progressing by short and repeated episodes

Pain in the gluteal regions and in the posterior part of the thighs,

Inflammatory involvement of peripheral joints, either p... articular and progressing in spurts, affecting only a few joir... only a single joint

Tendinitis

by pain which may be confined to this region of the spine or which may radiate to the thorax the patient becomes increasingly round shouldered, and the shoulders and the head project forward During this period, *involvement of the costovertebral joints causes the* amplitude of the thoracic motions to diminish, the thorax becomes increasingly flat antero-posteriorly, and respiration assumes principally the abdominal type

Soon or after many years, exacerbations of torticollis supervene Then the neck gradually becomes rigid and is often bent forward and turned slightly to one side, while the intensity of cervical pain tends to diminish As in the other spinal segments, when ankylosis has developed, the patient no longer suffers much from pain The cervical spine also having become ankylosed the patient is then obliged, in order to shift his visual field to move the entire trunk which now forms a single block with the head At that time the picture of Ankylosing Spondylitis is characteristic, and its diagnosis is evident

The *spinal evolution* of the disease varies greatly it may be continuous, it may be marked by exacerbations separated by long periods of remission, or it may even be latent

✓ *Inflammatory exacerbations in peripheral joints* may appear at any time during the evolution of spinal involvement often exacerbations in the spine and extremities coincide. When these exacerbations persist, they may end in stiffness or in true ankylosis. The joints of the lower extremities and shoulders are specially prone to be affected From a functional point of view these peripheral sites of the disease play a very important role a patient with dorso-lumbar ankylosis in a good posture may be able to perform a practically normal amount of physical labor a patient with chronic involvement or with ankylosis of the hips, knees and ankles becomes an invalid

Iritis usually featured by repeated exacerbations, is very characteristic of the disease, it may or may not accompany the articular exacerbations.

The effect of Ankylosing Spondylitis on the general condition of the patient may include emaciation, loss of appetite, slight fever and asthenia and these symptoms usually appear during the phase of onset or during exacerbations of the disease

therefore, to analyze the symptoms related to its spinal and peripheral manifestations

Outline of Clinical Examination which is Necessary to Treat and to Follow Cases of Ankylosing Spondylitis

J Forestier's experience of 20 years has taught him the great usefulness of a methodic clinical examination, and especially of one which includes measurements, these basic data having been established, it is much easier, during the course of the disease, to determine the degree of *aggravation or improvement*, and the effectiveness of treatment.

At the beginning of clinical observation we have made it a point to record the degree of *pain* as shown by the number of sedative capsules or pills taken each day, and of the functional symptoms, the facility or difficulty of performing usual acts (dressing, toilet, putting on shoes, walking, going up and down stairs changes in spinal posture and also changes in the patient s general condition temperature, weight, appetite)

Patient's Height. When the patient's height is measured with a scale, it determines his general posture and enables one to ascertain whether the degree of kyphosis is increasing or not

We inquire of each adult male patient his height at the time of his latest physical examination for the Army and by noting the difference between this measurement and our own, we can thus have some indication of the degree of spinal deformity

Vertical Deviation We apply this term to the distance which separates the occiput from the vertical plane, with the patient standing with heels and buttocks in contact with the wall, knees extended, and with the head held as much as possible in its natural position, and the patient's vision horizontal, that is, with the palpebral commissure and the upper insertion of the tragus on a horizontal line This distance is expressed in centimeters and serves as a basis of comparison This measurement, which is most important, permits one to determine the general posture of the spine and to measure, with mathematical precision, the progress of anterior inclination (kyphosis) of the patient.

When the patient also presents a *flexion deformity of the hips*, measurement of the degree of anterior deviation being made with

Examination of the Spine

Spine
Patient's height and development
Degree of forward projection of the head
Cervical Spine
Posture
Flexion
Extension
Rotation
Lateral flexion
Dorso-lumbar Spine
Dorsal posture
Lumbar posture
Distance from fingers to floor
Flexion
Extension
Lateral flexion
Respiratory amplitude
Pain on antero-posterior pressure on the chest
Pain on latero-lateral pressure on the chest
Painful Points
Cervical
Dorso-lumbar
Sacro-iliac

the knees straight, that is, with the thighs parallel to the wall, the entire trunk will bend still farther forward Thus the degree of anterior deviation is influenced not only by the spinal deformity, but also by the defective posture of the hips. In order to determine, in cases with hip involvement, the degree of anterior deviation caused only by the forward inclination of the spine, it is necessary not to attempt to correct the flexion of the knees and to have the patient maintain the dorsolumbar spine against the vertical plane

Measurement of the occiput to-floor distance, with the patient in the recumbent position is more difficult, subject to more causes of error, and does not correspond to the patient's posture in ordinary life.

Dorso-lumbar Region The posture of this region is indicated in a simple manner for example, 'increase in kyphosis beginning at L-1, with long or short radius , "flat back , ' disappearance of lordosis' ' scoliosis", ' trunk leaning to the right from

Mobility of the Dorso-lumbar Region The *fingers to-floor* distance (Fig 2 a), when the patient bends forward, with straight arms and knees, measures at the same time and in a satisfactory manner the degree of dorsolumbar flexion and the degree of pelvic rocking at the level of the hips When these joints are normal, the fingers-to-floor distance furnishes a satisfactory indication of spinal mobility

The determination of active lateral flexion reveals the degree of muscle contraction in the vertebral gutters in the concavity which is the reverse of what normally occurs (Bowstring sign, p 27)

The exact determination of painful points on pressure over the spinal and para spinal regions, and over the sacroiliac region, with the patient in the recumbent position, enables one to localize the foci of evolution This examination which should be repeated annually, gives a rather exact idea of the evolutionary status of the disease

Measurement of Thoracic Expansion This is measured with a flexible tape which is passed around the thorax at the level of the nipples [1] the difference between the thoracic circumference in expiration and in forced inspiration furnishes an accurate measurement of thoracic expansion In normal persons this measurement is equal to or greater than, 6 cm

A decrease in thoracic expansion measures the degree of involvement of the costovertebral and costotransverse articulations.

Pain on sudden pressure over the thorax, with the two hands laid flat, one at the side of the chest and one at the other side, or one hand in front of the chest and the other behind it indicates the evolutionary stage of the process in these articulations (J Forestier)

Antero-posterior pressure explores especially the upper level of the thorax from D 2 to D-8 lateral pressure explores only the lower level from D-8 to D-12

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[1] Because of the well-known and considerable variation in the level of the nipples, especially in women, a still more accurate measurement can be obtained by adopting as the level of measurement, the upper or lower border of the fifth rib in the mid-clavicular line

Cervical Region The amplitude of the antero-posterior movements is measured by the distance which separates the chin from the sternal notch in extreme flexion and extension Lateral flexion, as well as rotation to the right or left, is measured by means of a goniometer from the position of equilibrium

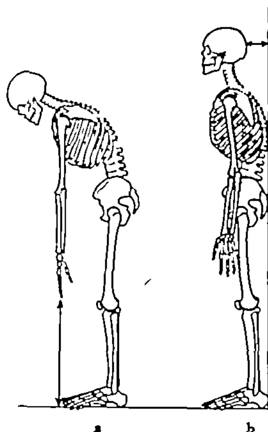


Figure 2. a. Measurement of the occiput to-wall distance which permits one to determine the spinal posture in the antero-posterior direction b Measurement of the finger to-floor distance which enables one to determine the degree of dorsolumbar mobility which may be modified by stiffness of the hips

The amplitude of these movements, which is regarded as normal for adult subjects, is as follows

Extension from the chin to the sternal notch (according to the length of the neck), about 18 cm ,

Flexion chin to sternum contact,

Right or left rotation 60° to 70°

Right or left lateral flexion 30° to 40°

Examination of Peripheral Joints

This examination especially as it concerns the lower extremities, must be meticulous

In cases with involvement of peripheral joints we employ an articular diagram inspired by Hans Jansen (Fig 3) The exam ination findings are indicated by means of conventional signs,[1] and the amplitude of the movements is noted in figures opposite each joint.

In cases with *involvement of one or both hips* it is necessary to measure systematically the principal passive movements as noted in the following table

RECORD OF AN EXAMINATION OF THE MOBILITY OF THE HIPS

Hips	*Right*	*Left*
Posture	Normal	Normal
Flexion	60 Pain.	75 Pain.
Flexion-Adduction	None	None
Abduction	30	30
Adduction	20	35
Rotation in extension	Diminished.	Normal.
Hyperextension	None—straight.	−5 from straight.
Turning in internal rotation	30	15
Turning in external rotation	10	15
Scarpa s pain point	+	—
Ischio-trochanteric pain point	—	—
Intermalleolar distance[2]	40 cm.	
Intercondylar distance[2]		

The figures given were taken from our records of a case with bilateral involvement of the hips.

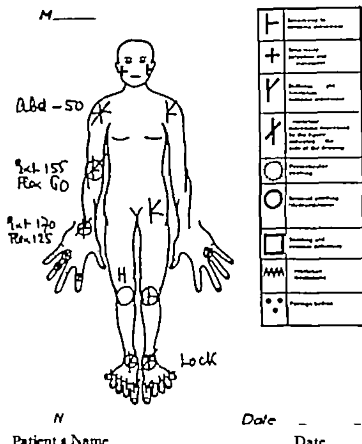
The degrees of motion are ascertained with the patient in the recumbent position on a hard bed first in the dorsal position and then in the prone position These degrees are measured from the position of equilibrium, that is, with the lower extremity prolong ing the axis of the trunk In this position of equilibrium, therefore, the thigh forms an angle of 180° with the trunk. In normal subjects flexion of the thigh diminishes this angle, the normal minimal limit of which is 35° to 40° When the thigh is in a position of flexion adduction, the knee can be brought level with the vertical line passing through the opposite lateral border of the trunk

[1] Pain is indicated by horizontal lines stiffness, by oblique lines and modifications of volume, by circles or squares.

[2] These measurements are important in cases of bilateral involvement.

The lower extremity can be deflected from the median axis of the body a minimum of 50° (abduction), and crosses this axis at a minimal angle of 40° (adduction)

Internal and external rotation are measured by the rotation maneuver of J Forestier, with the patient lying flat on the belly



Patient's Name Date

Figure 3 Articular diagram inspired by Hans Jansen this method of notation permits a rapid comparison of the condition of the peripheral joints at different dates of evolution.[1]

and with the knee flexed at a right angle, the position from which the degree of motion is measured is intermediate between internal and external rotations, that is, with the leg in a vertical position in extreme positions, that is, in full internal or external rotation, the angle formed by the *axis of the leg* and a vertical plane

[1] An explanation of this diagram (Fig 3) and method of notation will be found on the next page.

is about 35° Hyperextension of the hip is measured with the patient in the same position, normally this varies between 15° and 20° from the position of equilibrium.

This method of notation permits one rapidly to determine, by a simple comparison of diagrams, any modifications in the articular condition that may have occurred

The clinical examination is completed by a simple biologic examination, that is, by measuring the rate of sedimentation of the erythrocytes (Westergren method) and, if necessary, by roentgenography which enables one to determine the anatomic changes of the lesions.

In order to facilitate an easy comparison of the articular and general condition at different stages of evolution, the patients should be examined regularly every six or 12 months, and the different symptoms should be noted as succinctly as possible.

The intermalleolar distance is measured in dorsal decubitus, with the measuring tape between the two internal malleoli, with the two hips in a position of maximum abduction, and with the legs extended

The intercondylae distance is measured between the internal condyles of the two knees, which are bent at a right angle, and with the patient's heels touching each other This distance measures at the same time both the degree of abduction and of external rotation

These measurements are very important in cases of bilateral involvement of the hip

Spinal and Para-Spinal Symptoms

The symptoms in different parts of the spine, whatever may be their localization (lumbar, dorsal, or cervical), present certain common features which will be considered in groups before they are described topographically

Because of its anatomic and morpho-physiologic situation, we consider involvement of the sacroiliac joints as the first "spinal" symptom we will, therefore, study it immediately after the spine

The thoracic articulations, or more precisely the costovertebral articulations, are affected by the process at the same time as the articulations of the dorsal vertebrae, thus it has seemed wise to make a comparative analysis of the thoracic and spinal symptoms

SPINAL SYMPTOMS

Their General Character

Pain

Spontaneous Pain Although pain, which is a subjective manifestation, varies in each patient, it presents, in Ankylosing Spondylitis, certain features which very often permit one to suspect the diagnosis when the patient is first seen and questioned

Inflammatory Type of Pain This is dull and varies in intensity, with exacerbations and remissions, *it is in sharp contrast to the static type of pain* The improvement which the patient obtains from rest in the horizontal position is usually less definite than in the latter type of pain

Moreover, *night pain,* which disturbs the patient's sleep, often occurs Sometimes this kind of pain comes on at a fixed hour, between three and five o clock in the morning Patients say that this night pain may become so steady that they may be obliged to

rise several times, to take a few steps around the room or to take a sedative In the course of a bout some of our patients have had to spend one or several nights sitting in a chair

Night pain in the spine, which has just been described, may, with some variations, be observed in Pott s disease, in osteoporosis, and in unrecognized fractures of vertebrae, but Ankylosing Spondylitis is its most common cause. Without being absolutely pathognomonic, it represents one of the most suggestive symptoms of Ankylosing Spondylitis.

This feature of exaggeration by rest, or its appearance during rest, *is true not only for vertebral pain but also for* pain radiating to the posterior aspect of the lower extremities, for girdle pain, and for pain around the thorax

In the morning when the patient arises, his back is sore and the first movements are very painful and difficult The patient has the impression that his joints have become rusty Aspirin and other analgesics relieve the pain and thus help this morning "derusting' *which may require an hour or two*

The intensity of the pain varies greatly from a slight and quite bearable pain to pain which interferes with the patient's sleep and because of which he dreads the approach of night but this pain may be completely absent In a certain number of cases with complete spinal ankylosis it has been very difficult or impossible for the patients to recall any period of vertebral pain (see Latent Forms)

The evolutionary *rhythm* of the pain is variable At the beginning of the disease the pain is often intermittent and is often interrupted by short periods of relief later the pain takes the form of short spells over a continuous background

The pain is influenced by atmospheric changes, and especially by variations in humidity From a seasonal point of view, it may be much more severe towards the end of winter, during the second half of February, during the month of March, or during the first warm days of Spring A temperate and dry climate, on the contrary, tends to alleviate the pain

The intensity of the pain tends to diminish as the degree of stiffening increases *It may persist in the form of attenuated exacerba*tions in patients who preserve, either spontaneously or as the result of treatment, a considerable degree of mobility

Even when this spontaneous pain is the only symptom and when the sedimentation rate is normal, it must be regarded as a sign of active disease

STATIC PAIN Besides this inflammatory type of pain, there is a pain which is not due to the evolution of the inflammatory process itself, but which is caused by static disturbances in or around the spine and in the lower extremities In the cervical region, for example, the forward projection of the neck produces, in kyphotic patients, a marked contracture of the muscles of the neck, including the nape of the neck, and causes in the tendinous insertions of these muscles a pain which may be very troublesome (Trapezius Syndrome, p 74) The static origin of this pain is proved by the fact that, when the patients are straightened by orthopedic means, the pain disappears as soon as they have recovered a better posture

But in some cases the origin of the pain is more complex, static disturbances entail perispinal muscular contractures which not only tend to increase vicious postures, but which also tend to maintain the inflammatory condition As our experience has taught us, correction of the defective postures has a favorable action on the inflammatory condition, but basic treatment should not be neglected for this reason

Pain Induced by Spinal Examination PAIN ON ACTIVE MOBILIZATION Sometimes active movements of the spine cause pain which may at first account for their limitation but this pain is neither severe nor characteristic

PAIN ON PALPATION Most frequently this must be sought *at the level of the spinous processes or in the paraspinal zones* corresponding to the interapophyseal joints For this examination the patient should be as completely relaxed as possible he should be made to lie in the prone position either with the arms at right angles to the trunk or along the sides of the trunk

In patients' records which we have studied we have found spinal trigger points in only one half of the cases (53%-see Fig 4) Most frequently *multiple trigger points* are found these may be separated one from another, or they may be arranged in a more or less long chain which, in some cases, may extend over the entire spinal column, or nearly so The finding of disseminated pain points is an important element in favor of a diagnosis of Ankylosing

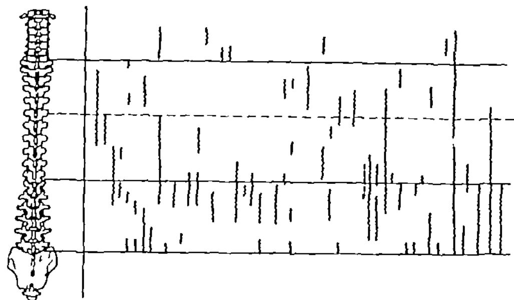


Figure 4 Diagram of pain points on palpation, showing their wide diffusion in Ankylosing Spondylitis (out of 100 cases of Ankylosing Spondylitis, 53 presented pain points, but only 7 patients had a single pain point, while in focal disturbances the pain was always localized)

Spondylitis, which from the beginning is a more or less diffuse disease

A single pain point was found in 7% of our cases. In Ankylosing Spondylitis it is important to be aware of this possibility In fact, the finding of a single pain point over the spine should not cause this diagnosis to be rejected and should not lead one to conclude that a focal disorder of the spine, such as Pott's disease, is present.

SACROILIAC TRIGGER POINTS These were found at the first examination in 8% of the cases, and they were present on one or both sides

ISCHIAL PAIN POINTS These points are sought by digital pressure over the tuberosity of the ischium, with the patient lying on his back and with the thigh semi flexed on the pelvis Then it is very easy to palpate the corresponding tuberosity The pain is most often elicited over the internal aspect. Having sought this sign during the past two years, we have noted its relative frequency without being able to define it accurately

PUBIC TRIGGER POINT Although, roentgenographically this joint rather frequently takes part in the inflammatory process, pain over this point was noted only once.

MUSCLES OF THE BACK CONTRACTURE—ATROPHY

The muscles in the posterior paravertebral gutters are often contractured, they are found hard, but of small volume, because the degree of atrophy which they undergo parallels the ankylosis of the spine (Fig 72b), and this accounts for the fact that muscular contracture is never very evident on exploration In destructive spinal processes (Pott's disease and spondylitis from other causes) we have observed an intense muscular contracture which does not permit any lateral motion and does not relax when, at the time of examination, the patient assumes the recumbent position

An already classical sign was described by J Forestier under the name of the "Bowstring Sign"

Bowstring Sign With the patient standing and with his back turned toward the examiner, he is asked to bend laterally to one side and then to the other

In normal subjects the muscles in the dorsolumbar (para vertebral) gutters become tense in the convexity formed by the bend of the spine, and they become relaxed in the concavity This phenomenon may be easily seen, the tense muscles being more prominent but it may also be felt since the same muscles give a sensation of firmness in contrast to the softness of the muscles on the concave side

In cases of early Ankylosing Spondylitis, even when spinal mobility does not appear to have diminished appreciably, the muscles in the lateral concavity of the spine contract, delicate palpation permits one to feel them under a tension greater than that of the muscles in the convexity of the spine

The phenomenon becomes especially perceptible when the patient is asked to *bend actively* during this maneuver, care must be taken that the patient moves exactly in the transverse plane without bending forward

We believe that this sign is due to the resistance which the muscles in the concavity encounter during the movement of lateral flexion, and which forces the patient into active contraction in order to carry it out

This sign is rather common, but to find it requires a certain finesse in technic Its presence is altogether characteristic

Relation Between Pain, Muscular Contracture, and Spinal Mobility The degree of muscular contracture is proportional to the degree of pain Pain plays a very important part in the reduction of mobility The fingers-to-floor distance (the method of following a case of Ankylosing Spondylitis, p 18), which gives an idea of the mobility of the dorsolumbar spine, is subject to very considerable variations in the same patient according to whether it is measured during an outbreak or during a remission Important gains in the degree of flexion of the spine should not be interpreted as due to improvement in the anatomic vertebral lesions, but simply as a consequence of the disappearance of pain and of muscular contracture

Involvement of the Lumbar Spine

Clinical Study

Lumbar involvement consists either in pain or in stiffness, accompanied or not accompanied by disturbances in spinal statics

Usually the lumbar spine is involved *at an early stage, but this is not necessarily the first symptom of the disease*

Lumbar involvement was the first symptom of the disease in 68 cases, and in 37 cases this involvement was limited to the lumbar segment of the spine, that is, without any other spinal or peripheral manifestation After the disease had progressed for one year, only half the cases presented lumbar symptoms During the first five years, lumbar involvement was present in 136 cases. This is to say that, at our first examination, which usually is after the disease has evolved from five to 10 years, this kind of involvement has been observed very frequently

Stiffness, without pain, had occurred in 35 cases, or in 17 5% of them. In these cases the diagnosis was usually made rather late the patients had not become aware of any definite manifestation in the region of the lumbar spine sometimes they had noticed some difficulty in performing certain movements, such as picking an object from the floor or ground But for many years this difficulty had not caused the patients any concern and thus, in 29 cases, the condition was discovered fortuitously in the course of an examination

TABLE OF LUMBAR INVOLVEMENT

Years of Evolution	*Percentage of Cases with Lumbar Involvement*
First symptom	34.0
1 year	51.5
5 years	68.0
10 years	82.5
15 years	88.0
20 years	95.5

Some of our patients showed lumbar involvement only after more than 20 years of evolution

Lumbar Pain Pain in this region has the same general character as other pains in Ankylosing Spondylitis *It progresses either by intermittent exacerbations*, and this is the usual type at the beginning, its intensity is usually moderate and it does not prevent the patient from carrying on his normal activities ("kinks in the back" and "muscular lumbago" are the names which are generally given to it) and this may continue for many years, or as a continuous lumbar pain, which may appear after an intermittent lumbago or which less frequently may be present from the beginning its intensity varies, and it may be very troublesome, especially when it occurs at night, as it not infrequently does.

This lumbar pain may continue for many years, with or without remissions. Pain of the inflammatory type tends gradually to diminish in severity and finally to disappear when lumbar ankylosis has become well established On the contrary, patients who retain good mobility often continue to suffer from more or less severe pain as long as they live

As we have already mentioned, pain may be absent from the clinical picture Stiffening then supervenes in a latent manner

Disturbances in Lumbar Statics In male patients lumbar lordosis disappears entirely or partly in half of the cases, and in a third of the female cases

In a few cases one may even observe, instead of lumbar lordosis, a dorsolumbar kyphosis, and nearly always this kyphosis has a large radius (Fig 30a) in these cases the kyphosis begins at the first or second lumbar vertebra

Limitation of Lumbar Mobility This limitation (Fig 5b) was observed at the first examination in 169 of our 200 cases or in

84 5% of them *it is, therefore, the most constant symptom of the disease* It was absent in only 15 5% of the cases these were cases in which peripheral articular involvement in the prevertebral stage (Modes of Onset, p 168) was present, in which transient and resolving vertebral exacerbations (Ill-defined Forms or Forms with Slow Evolu-

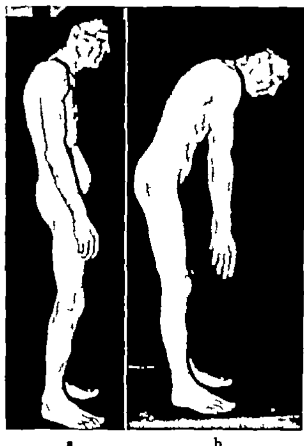


Figure 5 a. Ankylosing Spondylitis of 13 years' duration disappearance of lumbar lordosis, dorsal kyphosis, and forward projection of the head. b The finger to-floor distance permits one to appreciate the dorso-lumbar ankylosis which, in this case, is complete.

tion p 188) had occurred, and it included a few cases with a downward extension (Modes of Onset, p 173)

FEATURES OF THIS LIMITATION DURING THE EARLY PHASE Usually the movements are limited to the same degree in all directions, but sometimes certain movements are clearly more affected than others

According to our experience, limitation of the lateral movements of the spine is the earliest to occur this limitation is equal on both sides (in cases with protrusion of an intervertebral disc the limitation is greater on one side) To us the lateral movements appear to

be the first to be affected at least, these movements are the easiest to determine during an examination of the spinal column, in fact, when the patient's knees are well extended, the position of the pelvis during these lateral movements is fixed, while this is not strictly true during flexion and extension of the spine, when the pelvis can rock on its horizontal axis running through the hips Nevertheless, there are cases in which flexion is more clearly limited, and still fewer cases in which extension is limited

The limitation of movement often extends to other segments of the spine at the time of the first examination the dorsal region took part in this limitation in 47% of the cases Sometimes limitation of mobility in the dorsolumbar region was present, while the lower part of the lumbar region was free from it, this may have been due to the early appearance of bridging syndesmophytes at this level

The spinal column of a patient who is afflicted with Ankylosing Spondylitis gives, on inspection the impression of a *diffuse stiffening* or of a bar with little flexibility This impression is rather different from that which is produced by the spinal column of a patient with disease of one or more discs, in which, as long as movement is not painful, the spine remains supple This diffuse stiffening is also different from the marked but segmental, stiffness observed in Pott s disease

Ankylosis of the lumbar segment develops more or less rapidly

Roentgenology

First we will analyze the elementary roentgenographic lesions of Ankylosing Spondylitis bridging syndesmophytes, changes in the discs, in the vertebral bodies, and in the interapophyseal articulations. The changes in the lumbar region are described as the typical changes, and subsequently we will mention only the differences observed in the dorsal and cervical regions

After this analysis, we will describe the different aspects presented by the lumbar spine of a patient who is afflicted with Ankylosing Spondylitis

Basic Lesions SYNDESMOPHYTES (*From the Greek* συνδεσμοσ, *union, and* φυτοσ, *vegetation*)

J A Sicard and J Forestier in 1931, suggested the term "syn

desmophytes" for the roentgenographic images observed at the level of the discs in Ankylosing Spondylitis and compared these images with those observed in spinal osteoarthritis.

The definition which we would now give is as follows *the syndesmophyte is a bony formation that links two adjacent vertebrae* The situation and morphology of the syndesmophyte vary, it may insert itself more or less widely into the upper or lower third of the vertebral bodies and form a bridge in front of the disc space, or it may be situated at the outer part of a disc, where it may form only a moderate projection

In the first instance it seems to have developed from the deep, or subperispinal, layer of the fibrous sheath which surrounds the spine, or in the second instance it may have developed from the external fibers of the disc sometimes it develops from both of these anatomic structures (Pathologic Anatomy, p 271)

The appearance and distribution of syndesmophytes are characteristic of Ankylosing Spondylitis Only certain ossifications[1] resulting from an infectious or traumatic spondylitis may exceptionally resemble them, their presence solely at the level or in the vicinity of an abnormally thin disc, or of a deformed vertebral body, permits one to make the diagnosis.

Roentgenographic Study of a Fully Developed Syndesmophyte We have made roentgenograms of dry specimens (Figs. 6a and 6b) in order to avoid any superimposition of the images of soft parts, and thus to visualize the roentgenologic features with greater definition. We used a dry specimen in which the bony outgrowths were adjacent to the discs

In the *antero-posterior view* (Fig 6a) at the outer part of the disc, there may be seen a formation which links two vertebral bodies by means of a bridge This bridge is composed of three layers 1) a superficial cortical layer 2) a deep cortical layer, 3) and between the two a cancellous layer

The *superficial cortical layer*, which is very thin, describes a more or less salient convex curve and extends toward the lateral border of the body of the adjacent vertebra, where it unites with the cortical layer of the latter

[1] In secondary spinal neoplasms and in hydrofluoric intoxication (174) we find diffuse bony bridges opposite the discs, but in such cases characteristic and marked changes in bone structure are present.



Figures 6a and b a. Enlarged roentgenogram of lumbar syndesmophytes, seen from the front. The discs are confined by bony bridges inner cortical layer which is inserted into the marginal border outer cortical layer which is inserted into the middle part of the vertebral bodies between the two cortical layers is a layer of cancellous bone which is continuous with that of the vertebral bodies. b Lateral roentgenogram of syndesmophytes. These appear as thick laminae inserted into the marginal border



a

Figure 7a Photograph of a dry specimen of Ankylosing Spondylitis (lumbar segment of the spine, including L-1 L-2, L-3 and L-4) as seen from its right lateral aspect the bony bridges are voluminous and prominent they insert themselves into the lower or upper part of the vertebral bodies only the middle part of these bodies is free. It should be noted that the bony bridges do not encroach upon the intervertebral foramina.



b c

Figures 7b and c. b Photograph of a right median sagittal section of the same segment of the spine as that shown in Figure 7a the bony bridges do not bulge Ankylosis of the interapophyseal articulations should be noted c. Photograph of the same specimen sawed in the frontal plane (left half) the bony bridges which in the anterior part of the spinal column were composed only of a single layer of bone insert themselves into the angle of the vertebral body and at this point they present three layers a superficial cortical layer a deep cortical layer and a cancellous layer they insert themselves into the lower and upper thirds of the body This looks like a splitting of the single layer of bone visible on the anterior aspect of the bone.



Figure 8



Figure 8 bis



Figure 9

Figure 8 Mr Cart 50 years old, with Ankylosing Spondylitis of 10 years' duration early syndesmophytes at L-2 and L-3

Figure 8bis Mr Tri 27 years old, with Ankylosing Spondylitis of nine years' duration syndesmophytes with insertion into the vertebral angle of L-2 and extending vertically upward

Figure 9 Mr Fri 30 years old, with Ankylosing Spondylitis of three years duration early syndesmophytes at L-2 and L-3 (antero-posterior view) the lower bony formation is vertical and in contact with the upper third of the lower vertebra while the upper formation is inserted into the upper vertebra well above the vertebral angle.

The *deep, cortical layer*, which is rather thick, forms a vertical line and inserts itself into the angles of each vertebra. It is not visible at all angles

Between the two cortical layers there exists a cancellous layer which, at its more or less wide insertion, blends with the cancellous layer of the vertebral body sometimes vestiges of the original cortical layer of this body persist, and as a result the vertebral body seems to be widened at the level of these bridge abutments.

The appearance of the syndesmophyte is also clearly visible in roentgenograms made in the oblique position

Exceptionally in a lateral roentgenogram, the same image may be seen as in the antero-posterior view but usually the syndesmophyte is reduced to a single, thick, vertical line (Fig 6b) which recalls the inner cortical layer of the syndesmophyte as seen in a frontal view (Fig 6a)

The image just described as a complete bridge corresponds to an advanced stage of the syndesmophyte As the dry specimen shows, this appearance is due to a bony formation either outside the disc



a b

Figure 10



a b

Figure 11

Figures 10a and b Miss Wes , 24 years old with Ankylosing Spondylitis of 10 years duration Antero-posterior roentgenogram of the lumbar spine a. Syndesmophytes between L 3 and L-4 forming a complete but thin bridge b Roentgenogram of the same syndesmophytes three years later they have become much thicker

Figures 11a and b Mr Hyv 30 years old with Ankylosing Spondylitis of 10 years duration tomogram of a syndesmophyte made in the antero-posterior direction a. Section 9 cm from the dorsal plane the syndesmophyte is clearly inserted below the vertebral angle. b Section 8 cm from the dorsal plane, that is, farther back than the preceding one here the syndesmophyte is inserted throughout the upper third of the vertebral border and reaches the angle of the body

(outer cortical and cancellous layers), or at its outer limit (inner cortical layer)

Roentgenographic Study of the Development of the Syndesmophyte At the beginning and during the development of the syndesmophyte, it yields incomplete images in which one can see only one or both of the more or less developed piers of the bridge, but these have not yet joined

At its beginning, in antero-posterior roentgenograms, the syndesmophyte appears most frequently at the *lateral border of the vertebral body* at a distance of 3 to 5 mm from the vertebral angle, it presents itself in the form of a rough line, of which the starting point is fused to the vertebral body, it extends upward or downward vertically or slightly outward (Fig 8) The outline of this formation may be sharp, but *most often it is somewhat indefinite*

Sometimes it appears to start vertically from the vertebral angle (Fig 8bis) A curious, but rare, appearance may be produced by what might be called a non inserted syndesmophyte, which is separated from the vertebral body by a light, narrow cleft (Fig 9) The syndesmophyte may originate either from a single vertebra or from two adjacent vertebrae then the two formations progress towards one another and gradually become thicker, but this thickening of the syndesmophyte may occur only after the bridge has been established (Figs 10a and 10b)

As the syndesmophyte develops, its roentgenographic insertion point also widens and gradually invades the small space which separates it from the vertebral angle until this also is covered

In tomograms of a syndesmophyte we have observed that, in a section 9 cm from the dorsal plane (Fig 11a) where the syndesmophyte is less voluminous, it inserts itself below the vertebral angle which remains free In a section made 1 cm farther back, where the syndesmophyte is more voluminous, that is more fully developed it invades the vertebral angle (Fig 11b)

Very thick syndesmophytes may sometimes present a curious appearance for instance, that of a bagpipe (Fig 12)

Ten patients roentgenographed at different stages in the course of the disease have permitted us to determine the rate of development of a syndesmophyte This varies considerably, but in all these cases it took a minimum of two years for a bridge, when first seen at its very beginning to complete itself (Figs. 13a and 13b) this contradicts the assertions of some writers according to whom a syndesmophytic bridge may form in a few months [1] On the other hand cases occur in which during many years of evolution, syndesmophytes which had appeared at different levels have shown very little increase

It often happens that syndesmophytes of different degrees of development coexist on the same pair of vertebrae (Fig 14)

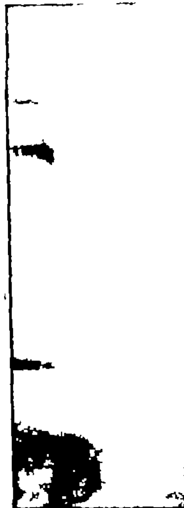
Once the bridge has been completed the deep cortical layer becomes more easily visible and all the elements described in the typical syndesmophyte can be perceived superficial cortical layer deep cortical layer and cancellous layer In roentgenograms the

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[1] On the contrary the formation of a syndesmophyte within a few months (Fig 17bis) is characteristic of the infectious forms of spondylitis staphylococcic, melitococcic.



Figure 12



a b

Figure 13

Figure 12. Mr Bru 47 years old with Ankylosing Spondylitis of 18 years duration thick syndesmophytes at L-2 and L-3 (antero-posterior view) with bagpipe appearance. The roentgenogram was well centered this appearance may be produced by a syndesmophyte that is only slightly salient, but which is viewed at a wrong incidence.

Figures 13a and b Mr Iso 35 years old, with Ankylosing Spondylitis of 10 years' duration antero-posterior roentgenogram of the lumbar spine. a. Syndesmophyte forming a complete bridge between D-11 and D-12 beginning syndesmophyte between D-12 and L-1 b Two years later the syndesmophyte between D-11 and D-12 has not changed but the syndesmophyte between D-12 and L-1 now forms a complete bridge



Figure 14 Mr Deb 45 years old, with Ankylosing Spondylitis of 18 years' duration the syndesmophytes have developed unequally at the edges of the same pair of vertebrae.



Figure 15 Syndesmophyte between L-2 and L-3 as seen in an antero-posterior view It is inserted into the lateral aspect of the upper third of the body near the vertebral border and extends vertically upward Its anterior and posterior borders can be distinguished thus bony formation hugging the contour of the vertebral plateau.

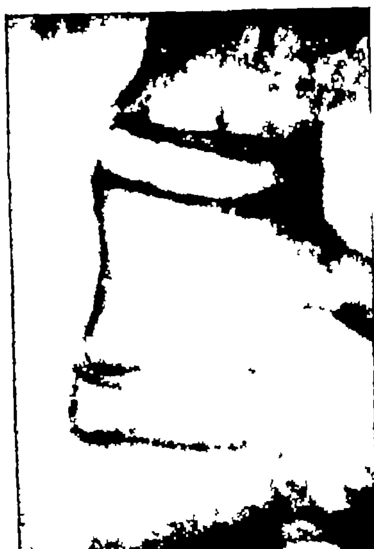


Figure 16 Mr Der 56 years old with Ankylosing Spondylitis of 34 years duration lateral roentgenogram of the lumbar spine thin syndesmophyte between L-2 and L-3 situated at the anterior aspect of the lumbar segment of the spinal column.

appearance of these different layers may not be simultaneous 1) most frequently the superficial cortical layer is the first visible element, and at the outset this manifests itself by a line or hook hugging the lateral aspect of the body a few millimeters from the vertebral angle 2) less frequently the first visible element is the deep cortical layer in the form of a vertical line inserted at the vertebral angle

Then the roentgenographic image gradually completes itself But during the entire course of the disease, although the two forma

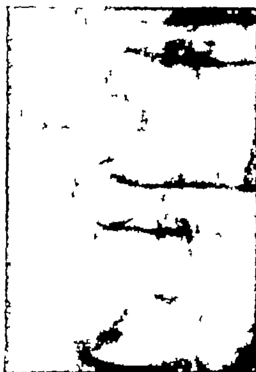


Figure 17 Mr Mell farmer 53 years old, with Ankylosing Spondylitis of 10 years' duration syndesmophyte developing on a pre-existing osteophyte (Fig 28a, b, and c)

tions exist (Fig 15), a single element may be visible in the films. However the deep cortical layer may, as we have already mentioned, constitute the syndesmophyte by itself, this is especially likely to occur at the anterior aspect of the spinal column and, in order to visualize it, a lateral roentgenogram of the spine is required (Fig 16)

Roentgenologic Differences Between Syndesmophytes and Osteophytes When the syndesmophyte begins to form, it does not always extend in a clearly vertical direction sometimes it may extend outward (Fig 17) It must be distinguished from the osteophyte which is observed in degenerative processes Nevertheless, such cases do not occur frequently and moreover, typical syndesmophytes may be seen at other levels in the same film (Figs. 28a, b and c) To us the more or less marked obliquity of the syndesmophyte in the zone of onset seems to be related to the previous condition of the discs In a case where previous degeneration of the disc has been present, the disc protrudes laterally and prevents the syndesmophyte from assuming the usual vertical direction and forces it to follow its contour Also in cases in which a degenerative process with osteophytes already exists, the syndesmophytes which develop

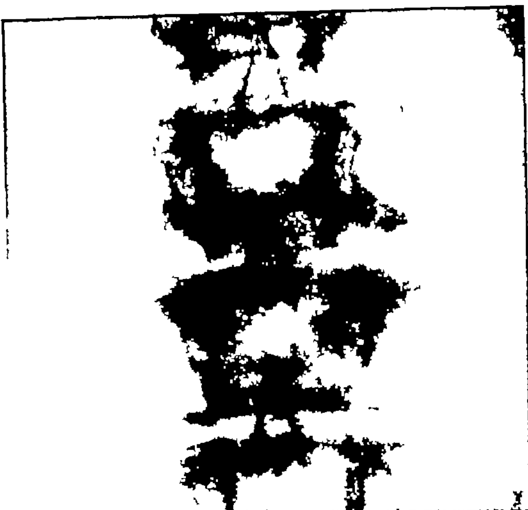


Figure 17bis. Mrs. Chi 71 years old record J F 12,591 Spinal pain with an abrupt onset temperature 40 C. for many weeks kept at rest on a hard bed gradual disappearance of the symptoms in three or four months walking resumed at the eighth month Successive roentgenograms show the appearance and evolution of a syndesmophyte at the level of the vertebral pair L-1 and L-2. In this film, which was made during the fifth month of the disease can be seen a voluminous, handle-shaped syndesmophyte with thinning of the disc between L-1 and L-2. Diagnosis of infectious spondylitis.

on these pre-existent formations will be still more difficult to identify However, we again insist that such cases are unusual

In the great majority of cases it is possible to distinguish a syndesmophyte from an osteophyte by the features listed in the following table, page 44

In a few cases their appearance may not be sufficiently definite to permit this distinction one must then take into account the condition of the other elements of the spinal column

A disc of normal height (thickness) favors the diagnosis of syndesmophyte

An abnormally thin disc, with a dense appearance of the contour of the vertebral plateau, favors the diagnosis of osteophyte

In cases where the distinction is impossible one must examine the other segments of the spinal column

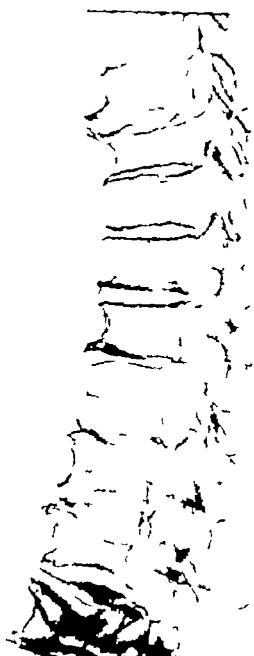
COMPARISON OF THE ROENTGENOLOGIC FEATURES OF THE SYNDESMOPHYTE AND THE OSTEOPHYTE

Roentgenographic Stage	*Syndesmophyte*	*Osteophyte* (Figs. 18a and b).
1 *At the beginning*		
Zone of Insertion	1 Almost always from 2 to 5 mm. from the vertebral angle 2. Sometimes at the vertebral angle (visible especially in lateral roentgenograms), 3 Sometimes independent of the vertebral body	1 At the limit of the vertebral angle, with a rather wide base from the start.
Direction	Vertical or slightly inclined, and crosses the intervertebral space	*Horizontal* increasing the width of the vertebral plateau.
Appearance	Fluffy or as a thin line	Always thick, with sharp contours, triangular and with a wide base.
2. *More Advanced Stage*		
Zone of Insertion	Wider over the entire upper or lower third of the vertebra	Widens little.
Direction.	Vertical	Horizontal and increases with the width of the vertebral plateau.
3 *Formation from the same pair of vertebrae*	Join one to the other	Usually remain parallel.
When they unite,	which is the rule, they form a bridge by extending one toward the other	which is rare they approach one another at a very acute angle.

THE INTERVERTEBRAL DISC *In principle* the intervertebral disc is of *normal thickness or height* This is a useful sign by which to recognize the syndesmophyte Nevertheless, in the lumbar region we have found in 17 9% of the cases, that is, rather frequently, definite examples of flattened discs

This thinning may be limited to one or two discs, or it may affect all the discs in this region (Ankylosing Form of Spinal Involvement, with painless evolution [233]) But this thinning of the discs is *not characteristic of Ankylosing Spondylitis* for example, it may be due either to degeneration of the discs or to Scheuermann's

a



b

Figure 18a and b a Dry specimen of spinal osteoarthritis, antero-posterior roentgenogram the discs are abnormally thin the osteophytes extend the width of the vertebral plateaux, and those belonging to the same pair of vertebrae have a tendency to remain parallel. b Same specimen as in Figure 18a lateral roentgenogram

disease, both of which conditions may co-exist with Ankylosing Spondylitis

In a few cases in which a disc has become abnormally thin its density may appear to have increased and this seems to be related to its ossification (Fig 19)

THE VERTEBRAL BODIES Generally the *form of the vertebral bodies* is but slightly modified however, since the syndesmophyte develops at the level of the upper or lower third of these bodies, the result is that, in roentgenograms *the plateaux of these bodies appear abnormally wide Consequently, the concavity of their lateral borders seems to be accentuated* in the image known as 'bamboo spine" the out ward prominences are due to the syndesmophytes which are 'sealed to the vertebral bodies, and the hollow parts correspond to the unaltered middle portions of the lateral surfaces of these bodies

Sometimes wedge shaped vertebrae may be found in the lumbar region, and especially in the dorsal region this tendency is made evident by lateral roentgenography

We have observed two *roentgenographic changes in vertebral structure* a *diffuse osteoporosis* of the vertebral bodies in one fourth of the cases this is most clearly visible in lateral roentgenograms we have found this especially at the beginning of the disease (when the disease had progressed between 5 and 10 years) Also it is often detected in bamboo spines Most frequently this osteoporosis con sists of a reduction in the density of the vertebral bodies which be come homogeneous and appear to lack structure In cases with a severe or very marked evolution the density of the vertebral bodies may be similar to that of the surrounding soft parts Then the contour of an affected vertebra, which is outlined by a fine line resembling a line drawn with a pencil, is more clearly visible (Fig 20)

Osteoporosis is likely to be found in the severe forms of the disease, whether the patient has been bedridden or not we interpret it as a *bad prognostic sign* Later during the advanced stage, osteo- porosis seems to occur less frequently

Oppenheimer (152) found some degree of osteoporosis of 50 cases of Ankylosing Spondylitis thi Oppenheimer s second stage of the disease w



Figure 19 Mr To 35 years old with Ankylosing Spondylitis of 15 years' duration the lumbar involvement was latent. Multiple and marked collapse of the discs, and absence of any syndesmophyte This roentgenogram gives the impression that the discs are partly ossified

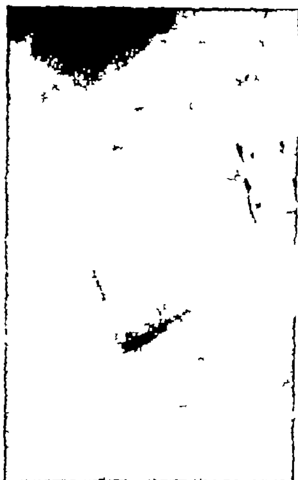


Fig 20 Mr Ver 21 years old, with Ankylosing Spondylitis of 10 years' duration lateral view of the lumbar spine diffuse osteoporosis of the vertebral bodies. Drops of *Lipiodol may be seen in the* intradural space. The clinical and roentgenographic symptoms presented by this patient were interpreted as due to an arachnoiditis, and he had undergone two unnecessary laminectomies.

75% of the cases, and it was found in 26% of the cases in the third stage.

Another very curious aspect may be found in the latent forms *the entire spinal column assumes a washed-out appearance* the structure of all the elements of the spine becomes crude as well as filamentous, these filaments, which are arranged vertically (Fig 27) diverge, at the level of the sacrum, toward the lower part of the sacroiliac joints

THE INTERAPOPHYSEAL ARTICULATIONS (188) In Ankylosing Spondylitis we have not systematically carried out a roentgenographic study of the interapophyseal joints because, from the stand point of early diagnosis,[1] the information obtained from oblique roentgenograms of these articulations is far less useful than that

[1] Since the majority of our patients were private patients, *it* was difficult to make a systematic study of joints which had only a meager diagnostic or therapeutic interest.

which is furnished by roentgenograms of the sacroiliac joints, or even than the information furnished by antero-posterior roentgenograms of the dorsolumbar region, which often show rather early syndesmophytes that are easy to identify

The features of these changes have been well described by A Oppenheimer (149, 152) Although we do not agree with him on the frequency and precocity with which they may be observed, we will now cite his description which, according to our findings, appears to be accurate

Oppenheimer considers three roentgenographic stages of these alterations

Stage 1, or Early Stage Mottled or diffuse *osteoporosis* of the articular apophyses, which varies from a slight reduction in density to an almost complete disappearance of the bony structure The edges of the articular facets are not sharp, and the articular space is somewhat thin or may even have disappeared

In one case the affected articulations had, after treatment, recovered their normal aspect, this seems to indicate that, at this stage, the abnormal changes may be reversible.

Stage 2, or Moderately Advanced The most significant sign is *erosion* roughness or waviness of the facets, and thinning of the articular space the osteoporosis is less definite (Fig 21) Sometimes, at the ends of the articular spaces, slight ossification may be found

Stage 3, or Terminal Stage This is characterized by bony *ankylosis* of the small interapophyseal articulations (Fig 22) this ankylosis is either complete, with disappearance of the joint spaces, or marginal, with persistence of the central portions of the apophyseal joints.

In Stage 1 the lesions are often limited to a small number of joints, while those of Stages 2 and 3 may be either local or general in all the interapophyseal articulations Usually not all of these joints present lesions in the same stage of development this occurs only during a very advanced phase The rate at which these lesions develop varies extremely

ROENTGENOGRAPHIC ASPECTS OF THE LUMBAR SPINE Quite different roentgenographic aspects of the lumbar spine may be observed These aspects are related either to different anatomic







Figure 21 Figure 22 Figure 23

Figure 21 Mr Ser 25 years old with Ankylosing Spondylitis of 14 years duration right interapophyseal articulation between L-3 and L-4 approximately normal articulation between L-4 and L-5 shows an irregular contour and osteoporosis of the apophyses.

Figure 22. Mrs. Cou 27 years old, with Ankylosing Spondylitis of 13 years' duration the right interapophyseal articulation between L-2 and L-3 shows an irregular contour and the right articulation between L-3 and L-4 is ankylosed An antero-posterior roentgenogram does not reveal any syndesmophyte.

Figure 23 Mr Rou 55 years old, with Ankylosing Spondylitis of 15 years duration marked sclerosis of the articular surfaces between L-4 and L-5 and between L 5 and S-1 on the left side.

types, to the soil in which Ankylosing Spondylitis had developed or evidently to the phase of its evolution

The lumbar spine may present an entirely normal roentgenographic image (Fig 24), and this occurs frequently at the outset.

Most frequently a single syndesmophyte or several of them are found, usually *in the dorsolumbar region* or in its vicinity (Fig 25) Often, also syndesmophytic images are found at several levels, but they are always most prominent in the dorsolumbar region

The 'bamboo' picture is rather rare and is observed only in a few very advanced cases. When it is present, syndesmophytes are found at all levels of the spinal column, and more or less voluminous bridges bind the vertebral bodies to one another Thus the lateral borders of the column form a continuous, wavy line, with bulges at the level of the syndesmophytes and hollows at the level of the

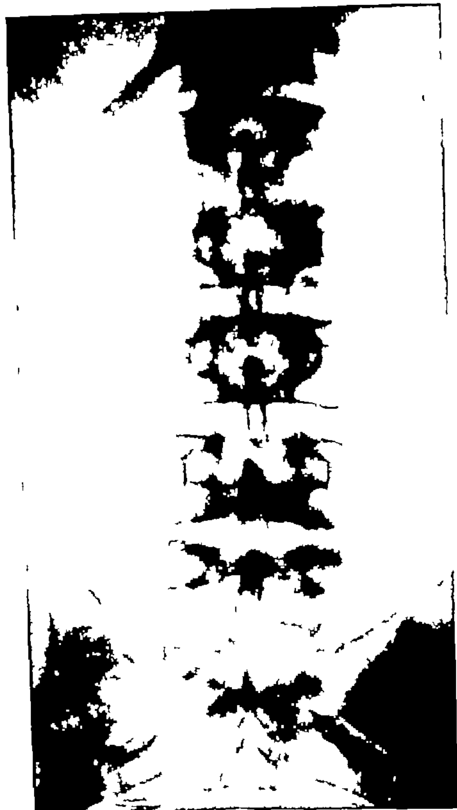


Figure 24 Mr Soren 24 years old, with Ankylosing Spondylitis of four years duration the patient complains of pain in the lumbar region Roentgenograms of this region do not reveal any lesion The sacroiliac joints show typical changes (Fig 63) as may be seen in this roentgenogram, the changes are difficult to analyze.

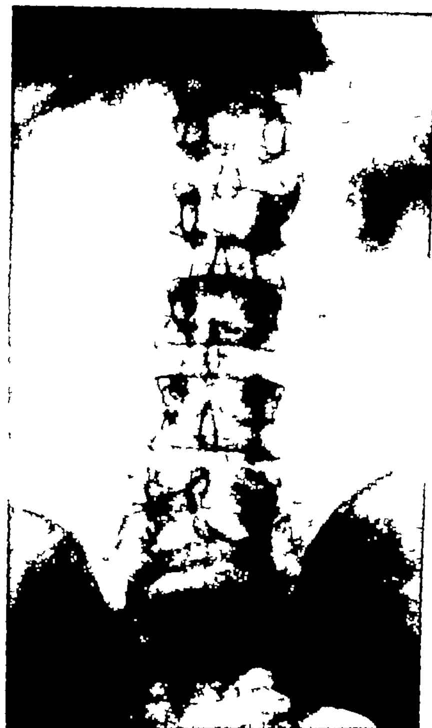


Figure 25 Mr Fonte 40 years old, with Ankylosing Spondylitis of 12 years duration antero-posterior roentgenogram of the lumbar spine. Multiple syndesmophytes are limited to the dorsolumbar region the sacroiliac changes are typical.

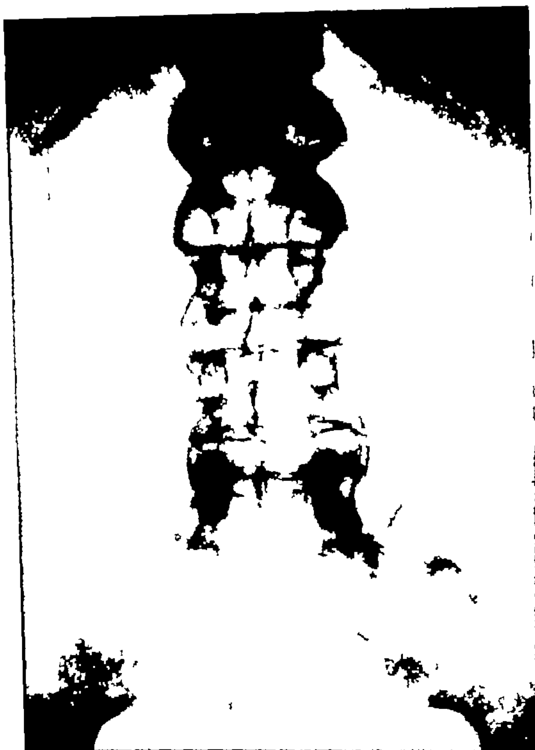


Figure 26 Same patient as in Figure 14 Picture of the "bamboo spine the changes shown are in an advanced stage. To be noted are the bulges and hollows created by the salient syndesmophytes

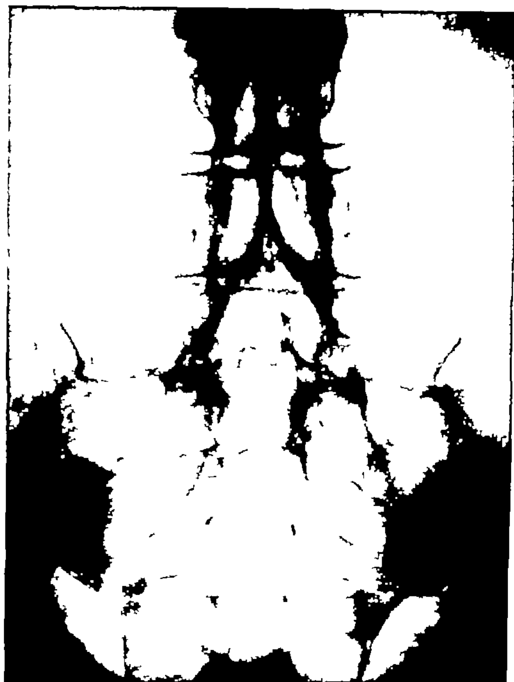


Figure 27 Mr Chev with Ankylosing Spondylitis of about 38 years' duration. A roentgenogram of the lumbar spine reveals a typical picture of the latent form thinning of discs, faint syndesmophytes, and three "rails" due to fusion of the interapophyseal articulations and to ossification of the interspinous ligaments.



Figure 27bis. Mr Chauf 41 years old with Ankylosing Spondylitis of about 16 years duration spinal ankylosis is established and the patient is free from pain Antero-posterior roentgenogram of the lumbar spine prominent syndesmophytes, discs of normal thickness, ossification of the interspinous ligaments and of the interapophyseal articulations. One has the impression that the discs are ossified this appearance might be produced by syndesmophytes on the ventral aspect of the spine. This picture is different from the two other films of the latent form which we have shown (Figs. 19 and 27)

bodies. The vertebrae and the discs themselves do not present any changes (Fig 26)

Another roentgenographic appearance, which also is uncommon, is that which we have described as most common in the latent forms (Fig 27) It is characterized by 1) three dense, parallel rails, the central rail being formed by an increased density of the spinous processes and of the ligaments that unite them the lateral rails being formed by the column of interapophyseal joints and of ligamenta flava very often the discs are abnormally thin 2) a washed out appearance of the vertebral bodies, and 3) some faint syndesmophytes which are not prominent (see Clinical Forms)

The three rails may occur in cases other than the latent forms, and they may be the only roentgenographic spinal sign



Figure 28a. Same patient as in Figure 17 Antero-posterior roentgenogram of the sacroiliac joints and of the lower part of the lumbar spine bilateral sacroiliac changes, most pronounced on the right side. Osteophyte between L-3 and L-4 typical syndesmophytes between L-4 and L-5 and between L-2 and L-3.

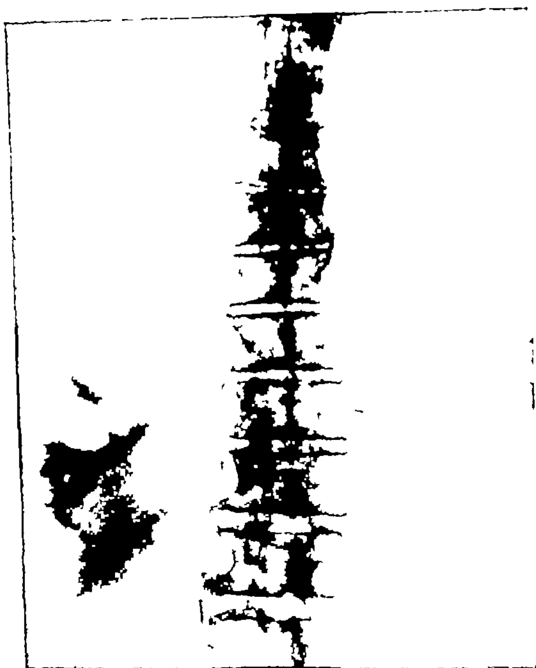


Figure 28b Same patient as in Figure 28a. Antero-posterior roentgenogram of the dorsolumbar spine typical syndesmophytes between L-1 and L-2 on the left side between D 11 and D 12, and between D-8 and D-9 syndesmophytes having developed on an osteophyte and producing complex images between L-1 and L-2 on the right, between D-12 and L-1 and between D-9 and D-10



Figure 28c. Same patient as in Figure 28a. Lateral roentgenogram of the dorsal spine. The syndesmophytes curve around the salient discs and, because of this, although the bridges are not thick, they bulge in front of the spine.

Another roentgenographic aspect may be observed when Ankylosing Spondylitis develops in a spine affected with hypertrophic arthritis or osteoarthritis in old persons who have done hard physical labor or who have been farmers. In the same film may be seen typical osteophytes, syndesmophytes and mixed syndesmophytic and osteophytic formations (Figs 28a, b, and c) For these forms some writers have proposed the name of *mixed* Ankylosing Spondylitis, a term which we do not use because it is not a question of a single, mixed process, but simply of the superimposition of *two different and successive processes*

In this region the first useful roentgenographic signs for diagnosis are the syndesmophytes, osteoporosis of the vertebral bodies and changes in the interapophyseal joints

The syndesmophytes are the most frequent and the earliest signs of lumbar involvement The earliest syndesmophyte which we have observed was found in a case in which the disease had been active only three years.

Osteoporosis of the vertebral bodies, which is always diffuse and especially visible in lateral roentgenograms, is often present, but when it occurs, it is, except in an occasional case, associated with syndesmophytes

Changes in the interapophyseal joints have seemed to us of little diagnostic value, roentgenograms, even when they are made in an oblique direction, are always difficult to interpret in relation to the contour of the surfaces and the condition of the joint spaces.

It must not be forgotten that, in Ankylosing Spondylitis, *the signs yielded by roentgenographic examination of the lumbar region are often negative until the disease has progressed for five years, and they are often slight until the disease has progressed for 10 years*

Nearly always the *first syndesmophytes* appear in the *dorsolumbar region*, and most frequently they affect the discs between D 11 and D-12, D-12 and L-1, L-1 and L-2, and rarely those between L-2 and L-3 In general roentgenograms of the lumbar region, when this region appears in the upper part of the films, there is danger of not being able to see syndesmophytes that are present Therefore, the films should be carefully centered on this region

There is no relation between the importance of clinical signs and that of the roentgenographic signs Cases with great pain may yield a normal roentgenogram latent cases, or more accurately 'painless'' cases, on the contrary may reveal pronounced roentgenographic changes

There is a certain relationship between the period of evolution and the stage of the lesions observed in roentgenograms In cases with syndesmophytes at different levels and forming complete bridges it is possible to assert that the disease has progressed for at least six years, and in cases in which *a bamboo spine is observed the disease has progressed for at least nine years*

Involvement of the Dorsal Spine

Involvement of the dorsal segment of the spine is featured either by *pain* with the same general character as in lumbar involvement,

or by *stiffness*, which may or may not be accompanied by an increase in kyphosis, which is a later symptom. When dorsal stiffness becomes established without pain, or when it is not accompanied by deformity, it often goes unnoticed on examination because of the difficulty of exploring the mobility of this segment.

Such involvement occurs later than in the lumbar region, which it usually follows in only 18 cases was it observed at the very beginning of the disease, and in practice it is never felt as a simple dorsalgia because it is always associated with lumbar or cervical symptoms, or with involvement of peripheral joints

TABLE OF DORSAL INVOLVEMENT

Years of Evolution	*Percentage of cases with dorsal involvement*
First symptom	9
1 year	16
5 years	39
10 years	57
15 years	68
20 years	73

At the level of the dorsal spine the most important problem is that of *alterations* in the spinal curve, and especially of an increase in normal kyphosis.

Kyphosis

This is the most common deformity from our statistics based on 200 cases, 136 or 68%, presented an exaggerated dorsal kyphosis its frequency varies according to sex 78% were in men, and 44% were in women We must recall the fact that, in the course of Ankylosing Spondylitis, lumbar lordosis disappears more often in men than in women.

The time of onset of kyphosis varies greatly moreover, when it has not been accompanied by pain it is difficult to determine accurately

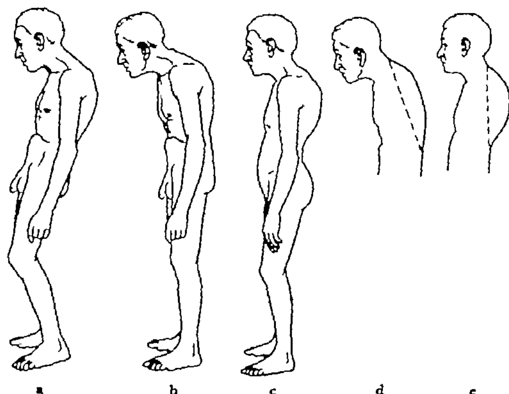
Description of the Type of Kyphosis Usually Observed in Ankylosing Spondylitis According to G Huc s definition, kyphosis is constituted by an exaggeration of flexion in front of



Figure 29 Mr Tur 47 years old with Ankylosing Spondylitis of 17 years duration lateral photograph high dorsal kyphosis and disappearance of lumbar lordosis for this the patient is obliged to compensate by bending the knees (also Fig 31a)

the spine or by transformation of the spinal curvature, which is normally concave behind, into a convex curvature

The patient bends forward with the shoulders projecting and the thorax flattened These cases of flattened lumbar segments, with the upper part of the back projecting forward and with the head and neck extending in a straight line with the axis of the back, give a family resemblance to the kyphosis present in all forms of Ankylosing Spondylitis These backs do not have a regular curve above and below its greatest prominence, as is true of other kyphotic backs here the kyphosis consists of a *lower, rectilinear segment* and of an upper segment which is bent forward Thus the kyphosis does not have a point of maximum deformity, or "summit,' but shows a zone from which the spine begins to bend forward and which is at the upper limit of the rectilinear, or vertical, segment



Figures 30a, b, c, d and e. a. Same patient as in Fig 29 Posture spontaneously assumed by the patient he compensates for the spinal deformity by flexion of the knees This posture permits him to look at distant objects. b Same patient as in Fig 29 the patient was asked to stretch his knees, when he could no longer keep his balance, the upper part of the body being drawn forward. c. Diagram of kyphosis of the same type as that encountered in Scheuermann's disease This is compensated for by lumbar hyperlordosis. d and e The "bowstrings" of the kyphotic curves (d kyphotic bowstring in Ankylosing Spondylitis e kyphotic bowstring in Scheuermann's disease) are if the different modes of compensation are taken into account, directed the first obliquely forward (forward inclination of the entire spine) and the second vertically In Ankylosing Spondylitis the most prominent point of the kyphosis is really its point of departure, while in Scheuermann's disease there is a point of greatest prominence, which is distinct from the starting point of kyphosis.

Lateral roentgenograms show that this forward inclination begins at different levels which we classify in two main groups

High Dorsal Kyphosis This type of kyphosis is by far the most frequent, the deformity beginning between D-4 and D-7 but most frequently between D-6 and D-7 (Fig 29)

Low Dorsal or Dorsolumbar Kyphosis The start of this deformity has been found between D 9 and L-3, and often between

L-1 and L-2 The kyphosis usually has a wide radius (Figs 31a, b, and c)

We will show the importance of this distinction in connection with the surgical treatment of kyphosis in Ankylosing Spondylitis There is not only an absence of balance by increased lumbar lordosis (a balance which occurs in kyphoses of the type found in Scheuermann s disease), but the normal lumbar lordosis also disappears Because the lower segment becomes stiffened, and because the spinal muscles undergo a considerable degree of atrophy, lumbar compensation cannot develop when kyphosis appears

As the center of gravity of the body tends to be displaced forward the patient tries to compensate for this loss of equilibrium by hyperextension of the hips but very often this hyperextension is no longer possible because the disappearance of lumbar lordosis has caused the pelvis to rock, and then compensation is accomplished by the knees which assume a more or less marked flexion (Figs 29, 30a, b)

Kypho-Lordosis In a few cases we have observed a simultaneous increase in dorsal kyphosis and lumbar lordosis This is typical of Scheuermann s disease (see Fig 30c) but it rarely occurs in Ankylosing Spondylitis In two cases of Ankylosing Spondylitis in which we have observed it we also found, in the lower part of the dorsal region, typical sequelae of Scheuermann s disease associated with lesions of Ankylosing Spondylitis which had developed subsequently

Roentgenographic Anatomy of Kyphosis As shown by lateral roentgenograms and dry specimens, this deformity is produced either by a posterior gaping of the disc (Fig 31d), by anterior thinning of the disc, or by a wedge-shaped deformity of one or more of the vertebral bodies (Figs 32 and 33), usually at the level of the middle or lower part of the dorsal region and of the upper part of the lumbar region These mechanisms may be associated in the same patient

We believe that *atrophy of the erector spinae muscles* plays an important part in the mechanism of these kyphoses This muscular atrophy is secondary to an inflammatory disease of the spine (arthritis of the apophyseal joints) The bodies and discs of this pathologic spine are malleable, their deformity, as well as the

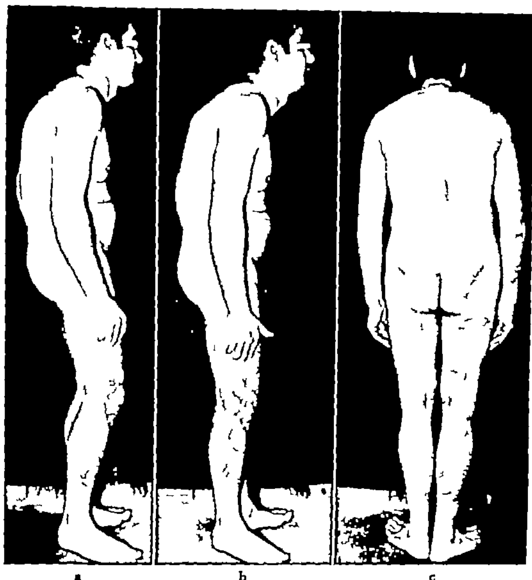


Figure 31a, b, and c Fon 32 years old with Ankylosing Spondylitis of 14 years' duration Dorsolumbar ankylosis and marked cervical stiffness dorsolumbar kyphosis with a wide radius. a. Side view showing spontaneous posture In order to maintain his balance and to look at a distance, the patient *tilts backwards and flexes his knees*. b. *Same patient. In order to maintain his balance when the knees are not flexed* all his muscles must be contracted Note the unflattened thorax and the prominent abdomen with supra-umbilical folds. c. Back view The bulges caused by the lumbar spinous processes are clearly visible. Note, in this patient, the good general appearance and the rather thick adipose tissue but examination reveals a distinct muscular atrophy



Figure 31d Same patient as in Fig 31a, b and c lateral roentgenogram of the dorsolumbar spine, in the erect position in this film posterior gaping of the disc between L-2 and L-3 is distinctly visible, and a similar though less wide, gaping of the disc between L-1 and L-2. These discs correspond to the starting point of the kyphosis.



Figure 32 Figure 33

Figure 32. Mr Sch 48 years old with Ankylosing Spondylitis of 15 years' duration lateral roentgenogram of the dorsal spine dorsal kyphosis due *to deformity of many vertebral bodies (wedge-shaped vertebrae) the discs are* of normal thickness.

Figure 33 Mrs. Eld 34 years old, with Ankylosing Spondylitis of 15 years' duration lateral roentgenogram of the dorsal spine high dorsal kyphosis, and slight wedge-shape of the body of D-6 When a film of the entire thorax was made, ankylosis of the manubrio-sternal articulation was revealed.

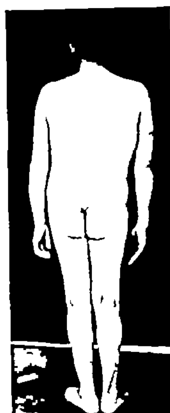


Figure 33bis. Mr C 37 years old with Ankylosing Spondylitis of 15 years duration Dorso-lumbar ankylosis, and marked cervical stiffness The trunk is bent forward and to the right this type of deformity may be found in one-fourth of the cases of Ankylosing Spondylitis Often these complex deformities are more marked than that shown in this photograph

tendency of the disease to stiffen the inflamed joints, rapidly set this form of kyphosis Compensation at the level of the lumbar region or of the hip joints is not possible because of pre-existing stiffness in the lower segment

The degree of kyphosis varies from slight deformity, which is not sufficient to cause the occiput to project forward of the dorsal plane, to a wide forward curve with flattening of the ankylosed thoracic cage Such a wide curve is quite embarrassing for the patient, while a dorsolumbar ankylosis in normal posture may go completely unperceived by the patient himself (Fig 34) it is the deformity, when it is marked, that causes embarrassment. *There is a distinct difference in the degree of kyphosis between men and women* out of 50 female cases only four had a forward inclination of the head equal to or greater than 10 cm (p 19—Vertical deviation), cases with marked kyphosis are exceptional To sum up in women forward deviation of the head is less common and usually slight (in general the deviation is less than 5 cm) in men on the contrary, forward deviation is more frequently greater than 5 cm and may reach very high figures.

Brief Study of Cases With Pronounced Kyphosis[1] (With forward deviation of the head greater than 15 cm.), based on 12 cases.

Patients occupations the occupations of seven required hard physical labor (farmers, butchers, and metallurgical workers)

Five patients had a sedentary occupation, but they worked all day in a sitting position, behind a desk

Age at the beginning of the disease quite variable.

Years of evolution at most between 10 and 20 years

Type of kyphosis in nine cases the kyphosis was dorsolumbar in three cases it was cervicodorsal, with the neck projecting forward and fixed (with the chin 7 8, and 9 cm from the manubrium sterni)

A loss of extension of the hips nearly always accompanies these marked kyphoses with lumbar or dorsolumbar starting point the backward tilting of the pelvis, the brim of the true pelvis tending to become horizontal because of the low kyphosis, causes the loss of hyperextension of the hips to appear even more obvious In a few cases in which a cervicodorsal deformity predominates, this loss of extension does not occur

The rate of sedimentation is always increased in spite of long standing disease In fact, it would seem that faulty postures tend to maintain the inflammatory process in the affected joints.

Other Variations in Spinal Posture Not all patients who are afflicted with Ankylosing Spondylitis present a kyphotic back 24 5% of the cases preserve a normal dorsolumbar curve, and 6 5 per cent have a *completely flat back*, like an ' ironing board , that is, *all the antero-posterior curves have disappeared (see Fig 71a)* This kind of back is most often encountered in women

This kind of flat back was seen in eight women out of 50, while it was seen in only three out of 100 men

It should be mentioned that these patients with flattened spinal curvatures had generally remained in bed a long time

LATERAL DEVIATIONS OF THE SPINAL COLUMN IN ANKYLOSING SPONDYLITIS Lateral deviations may be superimposed on the kyphosis and may aggravate the deformities (Fig 33bis) This is

[1] We have not seen any cases with neurologic disturbances related to these marked spinal deformities.





Figure 35

Figure 34

Figure 34 Mrs. Jou 47 years old, with Ankylosing Spondylitis of about 33 years' duration Dorsolumbar ankylosis had developed painlessly high dorsal kyphosis with anterior thinning of the discs continuous border line formed by the vertebral bodies and syndesmophytes in front of the discs.

Figure 35 Mr Etr 41 years old with Ankylosing Spondylitis of 29 years duration complete ankylosis of the spine deformed peripheral joints lateral roentgenogram of the dorsal spine kyphosis and thick syndesmophytes on the ventral aspect of the spine

to be observed in nearly one fourth of the cases a stiffened hip with loss of complete extension may be the origin of this deformity More often a defective posture establishes itself during a violent attack, in the course of which the patient has sought, when out of bed, a resting position one of our patients with a markedly deviated spine had passed through a very severe episode lying half on the side, in a so-called ' transatlantic type of chair and thus a spinal deformity in an "attitude of rest' had become established

A true scoliosis, with compensatory curvature and rotation of the bodies, is more rare, and it is always combined with kyphosis We have observed it in four cases

Very exceptionally one encounters a patient who presents a *pronounced and very complex kypho-scoliosis Of this we have* seen but two cases which may be called a malacic form of the disease.

Roentgenology In antero-posterior roentgenograms the *Syndesmophytes* first appear in the dorsolumbar region, and subsequently at other levels because of the superimposition of the costovertebral articulations, their interpretation is more difficult than in the lumbar region nevertheless, in the dorsal region they are quite visible

In lateral roentgenograms, according to the previous condition of the discs, the syndesmophytes alone or with the anterior borders of the vertebral bodies may present different aspects

A regular and continuous line borders the dorsal spine anteriorly (Fig 34) In a few very advanced cases the anterior part of the disc space is filled with bony tissue, while the thickness of its posterior portion is normal and the vertebral plateaux have remained parallel (Fig 35) Fusion of the vertebral bodies may also occur by thinning of the anterior part of the disc, this may occur in the zone where the kyphosis has started

This line is often slightly wavy because the syndesmophytes follow the curve of the altered and slightly prominent discs (Fig 36)

When the syndesmophytes have formed on pre-existing and marked osteophytic outgrowths, this waviness may be very pronounced (see Fig 28b)

At the anterior border of the spine a *pathologic image is often*



Figure 36 Mr Dou 45 years old with Ankylosing Spondylitis of nine years duration lateral roentgenogram of the dorsal spine kyphotic posture with a wavy bordering line.

lacking In this event the nature of the disease cannot be recognized by basing one s self solely on lateral roentgenograms

When the syndesmophytes are strictly limited to the right side of the dorsal spine, they must be distinguished from hyperostosis

In the chapter devoted to roentgenographic anatomy (p 63) *the deformities of the vertebral bodies* have been studied in relation to the roentgenologic anatomy of kyphosis *The condition of the discs* thinning of the anterior portion and replacement of the disc tissue by bony tissue have already been described

Recently attention has been drawn to the early changes in the *costovertebral joints* (219) Our impression is that they are not as early nor as useful from a diagnostic point of view as the author claims. As we have already seen, the dorsal region does not usually become involved until the disease has progressed for five or more years

In the dorsal spine osteoporosis and alterations in the interapophyseal articulations (Fig 37a and b), and in the costovertebral and costotransverse articulations, are difficult to interpret.

Involvement of the Cervical Spine

When this region becomes involved, the patient becomes aware of it at once, either because of the pain or because of the stiffness which even though it may not be painful is much more disturbing than when it occurs in other parts of the spine When the patient tries to raise or turn *his head*, *he feels pain at the back of the neck*, and he assumes a stiff posture later there supervene a true limitation of these movements and a progressive forward inclination of the head, which is so characteristic of Ankylosing Spondylitis then, in order to relieve the pain, most patients, when they go to bed, increase the thickness of their bolster or pillow

At the outset of the disease the neck is affected in only 7% of the cases (isolated or associated onset, p 173) In more than half of the cases this kind of involvement develops after the disease has progressed from four to 12 years, and it usually follows involvement of the lumbar and dorsal segments and it may appear *very late* we have observed cases in which cervical involvement had developed *only after the disease had progressed for 27, 28 and even* 34 years Since this kind of involvement does not develop so



Figure 37 Mr Delm 41 years old, with Ankylosing Spondylitis of 13 years' duration dorsal pain for several months, painful point on pressure over D-11 and D 12. a. Frontal view of the dorsal spine. b Lateral roentgenogram of the same region Numerous syndesmophytes, picture of bony destruction of the vertebral bodies D-10 and D 11 The disc between D-10 and D-11 is not abnormally thin because of ankylosis of the interapophyseal articulations (for the same reason, pain is not severe)

early as involvement of other parts of the spine, it is less frequently observed at the first examination

TABLE OF CERVICAL INVOLVEMENT

Years of Evolution	Percentage of Cases with Cervical Involvement
First Symptom	7%
1 year	13%
5 years	24%
10 years	36%
15 years	42%
20 years	52%
more than 20 years	66%

Clinical Symptoms

Cervical pain has the same general character as pain due to Ankylosing Spondylitis in any other region Torticollis, which is the usual manifestation progresses initially by more or less repeated bouts which are separated by complete remissions then it tends to become continuous, while stiffness gradually becomes established In some cases the stiffness may develop without pain

Posture The neck projects forward and usually forms a straight line with the dorsal kyphosis, without correcting it sometimes the neck may bend backward and thus may compensate more or less for the dorsal kyphosis by hyperlordosis (see Fig 31a) In cases with marked stiffness a slight lateral inclination of the head with slight rotation toward the opposite side, is often associated

Anterior displacement of the center of gravity of the head keeps the muscles at the nape of the neck, and particularly the Trapezius muscle under constant tension in order to support the head, and this tension ends by becoming painful (Fig 38) Thus there develops a syndrome similar to the syndrome described by Huc in other diseases under the name of "painful Trapezius syndrome" Pain of this kind which has a static origin should not be regarded as a symptom of progressing disease, but as a consequence of the defective posture which it entails.

Roentgenography permits one to recognize the postures in which the cervical spine has become stiffened

Here are the different possibilities which may occur



Figure 38 Same patient as in Figure 29 Painful contracture of the Trapezius muscle, caused by anterior projection of the head

1 In nearly half the cases lordosis is maintained or slightly modified, either in degree or in harmony

2 In a third of the cases the cervical spine is straight (Fig 39) the anterior borders of all the vertebral bodies form a straight line which may diverge more or less from the vertical, that is, which may lean forward more or less

3 The lordosis is sometimes increased, as if the patient had succeeded in overcorrecting the dorsal kyphosis (Fig 40)

4 More rarely the curve is inverted (Fig 41) the cervical spine has undergone kyphosis Of this we have observed five cases, sometimes with a regular curve and sometimes with a slightly angular shape resulting from a slight slipping of one or of several vertebrae. In one case there was a true spondylolisthesis at C-6, which required surgical fixation

Thus the cervical spine, like the other spinal segments, tends to bend forward

Cervical Mobility In order to study limitation of movement (normal mobility p 19), one must take into account the part played by pain and by muscular contracture, outside of the exacer



Figure 39



Figure 40

Figure 39 Mr Mah 41 years old with Ankylosing Spondylitis of 20 years' duration lateral roentgenogram of the cervical spine ankylosis in a straight line by fusion of the interapophyseal articulations, and faint anterior syndesmophytes.

Figure 40 Mr Mair 27 years old, with Ankylosing Spondylitis of 10 years' duration lateral roentgenogram of the cervical spine hyperlordosis correcting the dorsal kyphosis. To be noted are the involvement of the inter apophyseal articulations and the anterior syndesmophytes.

bations, that is, during painless periods, the movements are limited only by the anatomic changes.

There may be *slight limitation* especially of lateral movement and rotation which are the first to be affected or there may be *a considerable degree of limitation* in which all the movements are affected those which persist take place in the upper part of the spinal column (Fig 42) The greater part of the flexor and extensor movements take place in the sub-occipital joint, involvement of which occurs late in the process.

In cases in which only the occipito-atlantal joint preserves its movements we have observed a curious phenomenon When the

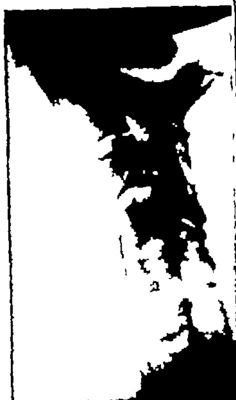


Figure 41



Figure 42

Figure 41 Mr Sav 57 years old with Ankylosing Spondylitis of 30 years duration lateral roentgenogram of the cervical spine slight kyphosis, thinning or obliteration of the interapophyseal joint spaces, osteoporosis of the vertebral bodies, and absence of syndesmophytes.

Figure 42. Mr Apov 48 years old, with Ankylosing Spondylitis of 17 years' duration lateral roentgenogram of the cervical spine ankylosis of the interapophyseal articulations, except at the level of C-1 and C-2, and of C-2 and C-3 continuous anterior line formed by the bodies and syndesmophytes which stop at the level of discs C-3 and C-4 Clinically the patient still has a certain freedom of neck movement, and especially of the antero-posterior movements.

patient is asked to turn his head to one side, he can do so only by simultaneously inclining the head toward the opposite side In other words, frank and pure rotation, without lateral inclination, is impossible

This is easily explained by the anatomy of the occipito-atlantal articulation and by the disposition of the lateral occipito-odontoid ligaments ' When the head and atlas turn toward the left, the right lateral occipito-odontoid ligament becomes tense If the

occipital insertion of this ligament could not be displaced, this tension would soon arrest the movement. When rotation continues, the ligament exerts traction on the right condyle of the occipital bone and the head inclines toward the right. When the ligaments are cut, rotation occurs without any lateral inclination. In living subjects this lateral inclination of the head is masked by an inverse curve of the cervical part of the column (182). In a patient whose cervical articulations, except the occipito-atlantal joint, have undergone ankylosis from Ankylosing Spondylitis, this compensatory movement cannot take place and lateral inclination of the head, which accompanies rotation, becomes evident.

In a few cases all motion is arrested and this is usually accompanied by ankylosis of all the other spinal segments. Then as far as movement is concerned, the head is a unit with the trunk. The field of vision of these patients will depend on the position in which the upper cervical vertebrae have become ankylosed. Moreover many cases with fracture of an ankylosed cervical spine have been reported; in these cases fracture was favored by the patient's inability to change the posture of the head at the time of the fall (224).

COMPARISON BETWEEN CERVICAL MOBILITY IN ANKYLOSING SPONDYLITIS AND IN CERVICAL HYPERTROPHIC ARTHRITIS OR OSTEOARTHRITIS. Measurement of the range of movement, in 25 cases of cervical hypertrophic arthritis or osteoarthritis, by the same method as in Ankylosing Spondylitis, showed preservation of the anteroposterior movements and moderate limitation of rotation and of lateral flexion. Never, save in a very painful case with cervico-brachial neuralgia, was there marked limitation or locking of motion similar to that which is observed in Ankylosing Spondylitis. Moreover the neck never showed the *faulty posture* and forward projection of the head which are observed in Ankylosing Spondylitis; in osteoarthritis cervical lordosis is often exaggerated.

ROENTGENOLOGY

Basic Lesions

The syndesmophytes are observed less often and later than interapophyseal joint changes; nevertheless, they remain the characteristic elements of the disease and their presence, even when they are

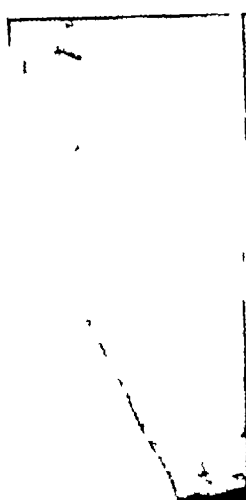


Figure 43

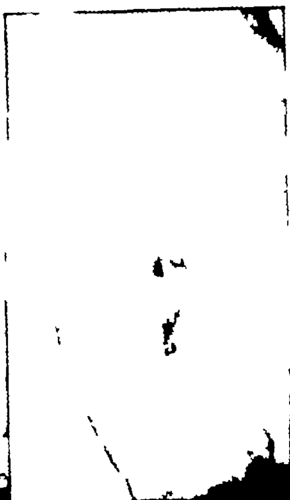


Figure 44

Figure 43 Mr Big 41 years old, with Ankylosing Spondylitis of 10 years' duration Lateral roentgenogram of the cervical spine, with ankylosis of the vertebrae in a straight line continuous and straight line formed by the anterior border of the vertebral bodies and syndesmophytes.

Figure 44 Mr Maz 45 years old with Ankylosing Spondylitis of 30 years duration lateral roentgenogram of the cervical spine the anterior syndesmophytes also cover the vertebral bodies and form a continuous and thick bony plaque which forms the anterior limit of the cervical spine. This peculiar appearance is found only in the cervical spine (dry specimens, Fig 130)

slight, permits one to make the diagnosis. Practically speaking, they are visible only in lateral roentgenograms and they often present appearances which are somewhat peculiar but which, when they reach an advanced stage, are typical

Then the feature most often observed consists of a more or less thin line which runs continuously in front of the cervical spine, and this line covers the vertebral bodies and the discs (Fig 43)

In extreme cases the anterior border of the spinal column is





Figure 45 Figure 46 Figure 47

Figure 45 Mr Bend Hospital 980 with Ankylosing Spondylitis of 15 years duration cervical involvement of five years' duration lateral roentgenogram of the cervical spine syndesmophyte in the shape of a hook inserted into the lower border of C-2 syndesmophytes of the same type, but much less pronounced, at the anterior and lower borders of C-3 and C-4

Figure 46 Miss Laan 24 years old with Ankylosing Spondylitis of five years' duration cervical spine involved 2½ years roentgenogram of the cervical spine syndesmophyte in the form of a handle with a rather indefinite contour

Figure 47 Mr Brai 48 years old with Ankylosing Spondylitis of 15 years' duration cervical spine involved for seven years lateral roentgenogram of the cervical spine syndesmophytes in the form of well-developed "handles."

formed by a thick and continuous lamina which covers the discs and bodies, the anterior limits of which can no longer be recognized But when the anterior part of the disc has become ossified, the thickness of the disc is normal (Fig 44)

In the course of its development the syndesmophyte may present features which are difficult to interpret

A simple thin, vertical line inserted into the vertebral angle, which may be distinguished from the osteophyte by its direction and often also by its hazy contours

A strong crook with a thick base and sharp contours, which is inserted into the vertebral angle (Fig 45)



Figure 48 Mr Saug 38 years old, with Ankylosing Spondylitis of 13 years duration lateral roentgenogram of the cervical spine ankylosis of the interapophyseal articulations, osteoporosis of the vertebral bodies, and faint syndesmophytes at the anterior part of the cervical spine

A strongly curved and fluffy hook, with somewhat indefinite contours, which is inserted into the vertebral angle or at some distance from it (Fig 46), and which, as it develops, produces an exuberant syndesmophyte in the form of a pot handle (Fig 47)

The Vertebral Bodies are not deformed (we have seen only one case with wedge-shaped bodies) In a few very advanced cases the bodies seem to be enlarged in an antero-posterior direction from the apposition of new bone (Fig 44)

The Discs are often abnormally thin throughout but even when their anterior portion has undergone ossification (Fig 44), they often preserve their normal thickness.

Changes in the Interapophyseal Joints develop early and they can be analyzed more easily than in other regions

Roentgenologic Appearances of the Cervical Spine

In the initial stage the articular zones appear to be osteoporotic while the vertebral bodies themselves may or may not show the same kind of change, at the same time the width of one or more joint spaces has diminished or they have become partly obliterated (Fig 41)

In the second stage is found complete ankylosis of the interapophyseal articulations which form a single block—so much so that the picture cast by these joints looks like a small, continuous column situated between the vertebral bodies in front, and the spinous processes behind (Fig 48)

In the more advanced forms the images are bordered anteriorly by the continuous line of the syndesmophytes, and posteriorly also by ossification of certain segments of the interspinous ligaments

In some of these advanced cases a variation of the picture is produced when all the bodies of the cervical vertebrae are fused into a single bone in which the discs and vertebral bodies can no longer be distinguished (Fig 49) It is noteworthy that even in these very advanced cases, oblique roentgenograms do not show any reduction in the caliber of the foramina (Fig 50)

Differential Diagnosis

This amounts to a recognition of the different causes of cervical stiffening

Cervical Involvement in Rheumatoid Arthritis Stiffness of the cervical spine is present in 20% of cases of this condition the tendency to increasing stiffness is much less pronounced than in Ankylosing Spondylitis. In roentgenographic films the joint spaces of the interapophyseal articulations are only narrower than normal syndesmophytes are not present A diagnosis can be made after an examination of the other spinal segments which in rheumatoid arthritis, are not involved



Figure 49



Figure 50

Figure 49 Mr Char 47 years old, with Ankylosing Spondylitis of 18 years duration lateral roentgenogram of the cervical spine it may be said that the cervical spine is transformed into a true long bone

Figure 50 Same patient as in Fig 49 This film of the cervical spine which was made with an oblique incidence shows that the foramina remain free in spite of the ossification of the other elements.

Considering the frequent roentgenologic changes in the interapophyseal joints in Ankylosing Spondylitis, the diagnosis should also be considered in relation to the forms of cervical osteoarthritis which are confined to these interapophyseal articulations, with minimal changes in the bodies and discs, and which may yield confusing roentgenologic images

We have studied eight cases in which osteoarthritis of the cervical interapophyseal joints was diffuse and very marked (Fig 51) In four of them this interapophyseal involvement was not accompanied by any changes in the discs or in the vertebral bodies (Fig 51)

The patients were more than 50 years of age five women and three men Except in one case, pain in the neck and shoulders had



Figure 51 Mrs. Crai 57 years old. Painful cervical syndrome for $1^1/_2$ years lateral roentgenogram of the cervical spine generalized and severe interapophyseal osteo-arthritis in the interapophyseal joints, with slight narrowing of the disc between C-6 and C-7

started recently The mobility which was rather well preserved was similar to that which we have described in classical osteo-arthritis of the cervical spine (p 78)

The diagnosis of osteoarthritis should be based on the following grounds

Recent onset in patients who are older than 50 years

The range of movement in the neck, which is less restricted than in Ankylosing Spondylitis

The absence of stiffness from the rest of the spine

The absence of inflammatory involvement from the peripheral joints

The increased density of the articular borders and the narrowing of the articular space are opposed to the osteoporosis and to the irregularity of contour and with still greater reason, to the ankylosis of the interapophyseal articulations Osteoporosis is an important element in the diagnosis of Ankylosing Spondylitis

PARA-SPINAL SYMPTOMS

Sacro-Iliac Symptoms (80)

In Ankylosing Spondylitis the sacroiliac changes are *constant* and they develop *early*, in order to orient the diagnosis from the very onset of the disease it is important to recognize their clinical manifestations

However, the existence of a clinical sacroiliac syndrome at the outset of the disease has not been realized or has been regarded as devoid of interest by the majority of writers who have thought it should be absent or faint, and that, when it is present, it should be associated with lumbar pain or with other early symptoms of the disease Thus, in 1942, Gilbert Scott (192) wrote that spondylo-arthritis in the phase of active sacro-iliitis is never painful Even *the absence of clinical signs* has been thought to be opposed to *the constancy of* roentgenologic signs in relation to these articulations

In 1945 however Boland and Present (19) mentioned some clinical sacroiliac signs at the outset of the disease and found these signs in about half of the cases Personally we have observed rather frequently a clinical sacroiliac syndrome consisting of pain, limping, and exploratory symptoms on examination

Clinical Study of the Sacro-iliac Syndrome

Spontaneous Pain It is localized at the very level of the affected articulation or it is referred to the buttock and to the posterior aspect of the thigh

The patient complains of pain, either in the lumbosacral region or in the gluteal region When he is asked to point a finger at the



Figure 51 Mrs. Cral 57 years old Painful cervical syndrome for $1^1/_2$ years lateral roentgenogram of the cervical spine generalized and severe interapophyseal osteoarthritis in the interapophyseal joints, with slight narrowing of the disc between C-6 and C-7

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site of the pain, he indicates an area over *the upper internal quadrant of the buttock* and usually this is bilateral

Present is a *deep pain with its seat in the buttock and in the posterior part of the thigh*, and more rarely along the internal or external aspect of the thigh, which may or may not be accompanied by pain in or about the joint itself Sometimes the pain extends as far as the popliteal fossa, but it never reaches the ankle or the foot *and it is* not accompanied by paresthesia

Character of the Pain It has the same general character as other pain caused by Ankylosing Spondylitis (see p 28), it is *deep, dull, moderate*, sometimes *lancinating*, and patients compare it with tooth ache

The improvement brought about by rest in the recumbent position is usually less definite than in vertebral pain of static origin the pain may even occur during the night and may interfere with the patient s sleep Sometimes it occurs at a fixed hour, towards three or four o clock in the morning

In the morning when the patient arises, he may be aware of soreness in the lumbosacral region Toward the end of the day the pain may reappear or may increase.

It should be mentioned that prolonged sitting may sometimes be definitely painful, the patient says that his chair is uncomfortable. He avoids remaining in this position for any length of time one of our patients was obliged to study in a reclining position

Limping Functional Disability Limping may be present on the more painful side

Most frequently pain and limping are slight and bearable, and the patient can carry on his activities but in some very painful cases the patients may have to use crutches and they may even have to go to bed for several months.

Pain and limping may be either intermittent and occurring in short bouts of from eight to 15 days, or they may be continuous with exacerbations.

Clinical Examination

Standing The patient presents a normal posture or he may show a slight *'list' toward the healthy side* he avoids resting his weight on the affected lower extremity which he holds with the knee slightly flexed



Figure 52. Internal surface of the Iliac bone (after Hovelacque) 1 Auricle or cartilaginous portion of the sacroiliac joint 2. Zone of insertion of the ligaments situated above and behind the auricle This zone is limited anteriorly by a rough line which extends upward the anterior border of the auricle 3 Postero-superior iliac spine 4 Inter spinous notch 5 Postero-inferior iliac spine 6 Pre auricular furrow 7 Innominate line

WALKING *On the affected side he may have a slight limp*, and climbing stairs may make this sign more apparent

INSPECTION The examiner may often find atrophy of one or both buttocks, which then stand out with less relief In one case with unilateral involvement of the sacroiliac joint the muscular atrophy also was definitely unilateral

PALPATION (In order to obtain good relaxation of the muscular masses this is always practiced with the patient lying prone), we seek pain by digital pressure on the posterior part of the joint, which is situated *immediately below the postero-superior iliac spine*, at the level of the third sacral foramen This is often sought at a level which is too high and adjacent to the lumbosacral joint

The sacroiliac joint is covered behind by the iliac crest which overlaps it posteriorly and internally, and also by the thick iliosacral ligaments Only the posterior horizontal extremity of the auricle which is situated below the postero-superior iliac spine, is accessible to the examiner's finger (Fig 52) Practically we have found this trigger point in disturbances of the sacroiliac joint, and it is the only one which, to us, seems reliable, because it permits direct examination of the joint Sometimes a very thick fatty pannicle may interfere with this examination

The many other neighboring *painful points, which are so often* found in this vicinity, do not permit one to examine the sacroiliac joint space, and to us they seem much more difficult to interpret

The maneuvers employed to induce pain on mobilization of the sacroiliac joints may be positive. These maneuvers are *numerous* and *difficult* to interpret *not one of them is pathognomonic* moreover there is often a *certain discordance between them*

In order to interpret the results of all these maneuvers, one must always keep in mind the following points when one mobilizes the sacroiliac joint, the corresponding hip and the lumbosacral region also are often mobilized (Fig 53) Therefore, the patient must be asked to point to the precise spot where the induced pain is felt. This maneuver may be considered to yield a positive result only when the patient has indicated the sacroiliac region

These maneuvers are employed to mobilize the sacroiliac joints either like scissors, in the antero-posterior direction, or by opening (separating) their anterior or posterior portion

MANEUVERS TO MOBILIZE THE SACROILIAC JOINT LIKE SCISSORS. a) With the patient in the recumbent position, one thigh is overflexed on the pelvis, while the other thigh is kept in the extended position b) With the patient in the prone (ventral) position one thigh is overextended on the pelvis with the knee flexed while the sacrum is firmly held with one hand c) Gaenslen's sign the patient lies on the side with the sacroiliac joint to be examined farthest from the table The lower extremity which rests on the table, is subjected to maximal flexion, the patient holding his knee with both hands The knee of the other lower extremity having been flexed the examining physician grasps it above the knee-cap with the opposite hand, he then places the other hand at the level of the gluteal fold and overextends the thigh on the pelvis d) A variation of this maneuver may be carried out with the patient lying on his back and with the lower extremity on the side to be examined extending beyond the side of the bed after this, the maneuver is identical to the previous one

MANEUVERS INTENDED TO OPEN THE SACROILIAC JOINT (either at its anterior or posterior part) a) With the patient lying on his back, the lower iliac spines are pulled apart, and thus traction is made on the anterior articular space (Ericksen) b) With the patient

in the same position, the iliac wings are forcibly brought together, and thus traction is made on the posterior part of the joint space c) With the patient on his back, and with the knee and hip on the side to be examined in flexion abduction, the foot remaining on the bed the anterior superior iliac spine on the opposite side is fixed with one hand and with the other hand pressure is made toward the plane of the bed on the knee of the side to be examined Thus the abductors are placed under tension at the same time that the sacroiliac joint is mobilized (Laguerre) d) With the patient in dorsal decubitus, the knee on the side to be examined is flexed and the external malleolus is placed on the opposite knee While the iliac crest on the same side is held in this position pressure is made on the knee of the side being examined

Rather often in the sacroiliac syndrome of Ankylosing Spondylitis, these maneuvers have yielded positive results Many other examination maneuvers have been described but in order the better to appreciate their value and their interpretation, it is wiser to carry out only the simpler maneuvers and always the same ones

SEARCH FOR NEUROLOGIC SIGNS These pains of sacroiliac origin are not accompanied on examination, by *objective neurologic signs* a) Superficial sensory disturbances are not present, b) The patellar and Achilles reflexes are normal c) There is no motor deficiency

An examination for Lasègue's sign may induce pain in the posterior part of the thigh, but this appears only when the lower extremity is strongly flexed on the pelvis This induced pain may be due either to pull on the hamstring muscles (we have often found the ischium painful on palpation and showing roentgenologic changes) or to mobilization of the sacroiliac joint

This mechanism may be brought out by Goldthwait's procedure With the patient lying on his back, one hand is slid beneath the lumbar curve with the other hand the leg is slowly raised, as if to test for Lasègue's sign When the patient complains of pain before the lumbosacral region has budged—the examiner will know this by the hand placed at this level—the pain will be due to pulling of the sciatic nerve, when the pain appears only at the moment the lumbar region is moved it may be due to mobilization of the sacroiliac joint

Through what mechanism does pain develop in the buttock and in the posterior part of the thigh when the sacroiliac joint is involved?

The classic explanation is that the nerve trunks of the lumbosacral plexus are irritated as they pass in front of the inflamed sacroiliac joint and this irritation of nerve trunks would also cause the pain in the posterior part of the thigh

But this explanation is far from satisfactory We know that a nerve trunk is made up of motor fibers and of superficial and deep sensory fibers stimulation of this trunk should affect all three elements, and thus besides the deep pain, motor as well as superficial sensory disturbances (paresthesia, hypo-esthesia or hyperesthesia) should appear in the territory corresponding to the affected trunk

But in the sacroiliac syndrome of Ankylosing Spondylitis, with pain in the posterior part of the thigh only a deep pain is observed in all the cases, therefore signs to indicate involvement of the superficial sensory fibers or of the motor fibers are lacking Thus it is impossible to consider pain of this kind as due to involvement of the nerve trunk or roots

The studies of Lewis and Kellgren (44) on the physio-pathology of pain have shown that, when pain that has originated in superficial structures is accurately located by the patients, pain that has originated in deep organs, such as some of the joints or viscera, is much less accurately localized and diffuse and it is often referred to the territory corresponding to the medullary segment that receives the fibers conveying sensory impulses from the affected organ (the segment S-2 for the sacroiliac)

We believe we can classify with this type of *deep referred pain with segmental distribution* the pain in the buttock and in the posterior part of the thigh of the sacroiliac syndrome in Ankylosing Spondylitis. In rheumatologic practice examples of this deep referred pain are known for example, pain in the anterior part of the thigh and in the knee in diseases of the hip

In practice just as pain in the anterior part of the thigh, without superficial sensory or reflex disturbances, directs one's attention toward disease of the hip so pain in the buttock and in the posterior part of the thigh, without neurologic disturbances, should make one think of the sacroiliac joint

The movements of the spinal column are free, it is only in very painful cases that a certain tension in the lumbosacral muscle masses may be noted, and this may result in a slight limitation of movement in this region In these cases, also pressure on the spinous process of L-5 may cause pain

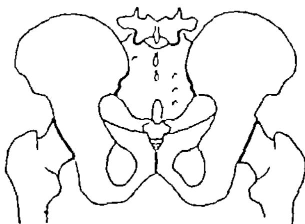


Figure 53 Diagram of the pelvis, the sacroiliac joints, the interapophyseal articulations L-5 and S-1 and the hips.

In brief, the symptoms which should make one think of sacroiliac involvement are a) Pain complained of by the patient at the level of the joint or around the buttock

b) Pain in the posterior or outer part of the thigh, without any neurologic signs to suggest that nerve trunks or roots are affected

c) The fact that the patient suffers while in a sitting position

But abnormalities in neighboring organs may cause pain in the same regions and may thus lead to confusion (Fig 53) Therefore, a *topographic diagnosis* must be made

DISORDERS OF THE LUMBAR AND SACROILIAC REGIONS Mechanical, inflammatory or neoplasmic disorders in the lower part of the lumbar region may cause pain in the lumbosacral region or radiating to the lower extremity and may thus raise a problem of differential diagnosis from sacroiliac involvement

Lumbo-Sacral Pain In principle the seat of spontaneous pain is higher than in the case of sacroiliac pain, but especially in the case of disorders of the lumbosacral disc, cases are encountered in which questioning the patient does not permit one to distinguish sacroiliac pain from lumbosacral pain

Lateral flexion deformity of the spine, with definite deviation of the trunk toward the right or left, and without a compensatory curvature, always favors protrusion of a disc and this lateral flexion deformity is different from the "hippy" posture that may be encountered in the sacroiliac syndrome, which is always slight and by means of which the spinal column compensates for the loss of equilibrium of the pelvis

Lumbar stiffness When it is present, it always favors a disorder in this region

On palpation one may find definite painful points over the spinous processes But a painful point over the spinous process of the 5th lumbar vertebra, or near it, may also be noted in sacroiliac disorders.

Maneuvers intended to mobilize chiefly the lumbosacral region, and carried out with the patient standing or sitting are likely to stir up lumbar pain On the contrary maneuvers intended to mobilize the sacroiliac joints, excluding movement in the lumbar region (Mennell, Ericksen, Laguerre, and Gaenslen) are negative in such cases.

Remote Pain Pain radiating to the lower extremities, when distributed in the form of a band and when accompanied by neurologic symptoms (paresthesia, superficial sensory disturbances, abolition of tendon reflexes, or motor deficiency), has, in principle, a radicular origin and nearly always a vertebral origin, and is most often due to protrusion of a disc (de Sèze [193])

Sacroiliac pain, on the contrary is a deep segmental pain that is referred to the posterior part of the thigh, extending to the knee or calf but *never reaching the dorsum of the foot or the great toe*, as does a radicular pain in L-5 nor does it reach the sole of the foot, as does pain in S-1 It is not accompanied by neurologic symptoms

A more difficult problem is raised in cases where a root pain extends only to the buttock or to the posterior aspect of the thigh, without reaching the foot But rarely, even in these cases are neurologic signs (paresthesia, etc) completely absent. It is then that spinal examination has its greatest value and is needed to orient the diagnosis.

In certain cases roentgenology will only serve to confirm the diagnosis of lumbar lumbosacral or sacroiliac involvement. But

in other cases where the clinical diagnosis may remain uncertain, well-centered roentgenograms should systematically be requested one over the disc between L-4 and L-5 and the sacroiliac joints, and the other over the disc between L-5 and S-1

DISORDERS OF THE HIP A mistake that is frequently made is to ascribe to the hip joint symptoms that have their origin in sacroiliac involvement The opposite error, that of attributing to the sacroiliac joint signs which are related to the hip joint, is less common

This confusion is explainable because, in both syndromes, there may occur

Pain on standing or when walking

The 'hippy' posture

Limping

Disorders of the hip are accompanied by pain in the middle and lower portions of the buttock This pain may be difficult to distinguish from that which occurs in sacroiliac disease But pain from the hip is accompanied by pain in the groin, and by deep pain referred to the anterior aspect of the thigh as far as the knee. This anterior femoral pain always points toward hip disease

It may be recalled that most of the examination maneuvers for the sacroiliac joints are the same as those for the hips, and that maneuvers intended to mobilize the hip also mobilize the sacroiliac joints, especially when they are carried to an extreme degree, and then they cause pain which is difficult to localize (this is one of the causes of diagnostic error which we have mentioned) The best diagnostic sign of hip involvement is limitation of passive joint movement In these cases one should practice the maneuvers with great care so as to avoid moving the pelvis this can be performed with the patient in the prone position, one hand holding down the pelvis To us the maneuver known as the "ball bearing test (gentle rotation) of the hip joint (J Forestier) seems to permit one to study the hip to the best advantage without mobilizing the sacroiliac joint

In doubtful clinical cases it will be necessary to take, not one roentgenogram centered on a single hip but to take a roentgenogram of the entire pelvis thus besides images of the hips, a fairly well centered image of the sacroiliac joints will be obtained

DISEASES OF THE SOFT PARTS IN THE LUMBOSACRAL REGION AND

IN THE BUTTOCKS Pain around the buttock may be due to disease of the soft parts, either subcutaneous (adiposalgia, fat herniation of Copeman and Ackerman [44]), or in the deeper tissues (muscles and perimuscular fibrous sheaths)

Cellulalgia is observed in obese women of a certain age It is easily recognizable on examination it is rarely localized, the skin is thick, adherent, and hard, and palpation of this skin, when it is gathered into rolls, causes a diffuse and sharp pain. The question of differential diagnosis between it and the sacroiliac syndrome seldom arises

Fat Herniation of Copeman and Ackerman Sometimes one may observe in the lumboiliac space, in the vicinity of the sacroiliac joints, nodules that Copeman and Ackerman have described under the name of fat herniations but in our experience they are only rarely painful on pressure, and to us they have not appeared to be responsible for a painful clinical syndrome

Frequency of the Sacroiliac Syndrome We have found clinical signs of sacroiliac involvement in 94 out of 200 cases, or in nearly half the cases.

In about a third of the cases in which sacroiliac signs were present *the syndrome was complete* pain around the joint and pain referred to the thigh In the remaining two-thirds the pain was confined to the joint, or *more frequently to the buttock or to the posterior part of the lower extremity*

In the other half of the cases the sacroiliac involvement was latent and did not produce any symptoms, either when the history was recorded or when the patient was examined

Time of Appearance of the Syndrome When this syndrome is present, it is usually discovered during the *phase of onset* of the disease.

It may be recalled that the *phase of onset* of Ankylosing Spondylitis is that during which a definite and permanent limitation of spinal mobility has not yet developed During this phase of onset, which often extends over many years, the disease usually manifests itself by intermittent progress, with remissions.

In 34 cases this was the first sign of onset and it constituted the entire clinical picture

In 31 cases it was *one of the signs of onset but it was associated with*

other symptoms of the disease, the clinical picture of which was more complex

In 29 cases, in which *the disease had manifested itself by peripheral signs, the sacroiliac syndrome supervened later*

Evolution of the Clinical Syndrome At the outset it may be *unilateral* (8) It may extend to the other side and become *bilateral*, either alternating from one side to the other, or it may occur on both sides simultaneously, in other cases it may evolve *bilaterally from the beginning* These sacroiliac episodes may persist by themselves for many years, without any other symptoms, and their progress may be interrupted by short or long remissions. In seven of our cases the clinical sacroiliac involvement was unilateral at first, and then it became bilateral

At some period of its evolution, either at the very beginning or later in its course, *lumbar or other symptoms* develop and the clinical picture becomes more complex.

This subsequent evolution toward a more complete picture of Ankylosing Spondylitis—a fact which has been verified in all the cases that we are reporting—permits us to state that this inflammatory sacroiliac syndrome truly belongs to Ankylosing Spondylitis. This is not well known, and the sacroiliac syndromes of Ankylosing Spondylitis, without spinal involvement, are usually erroneously diagnosed as sacro-coxalgia, sacroiliac osteochondritis, puzzling changes (1), and they are often wrongly fused on this basis (37a)

Roentgenology

For the diagnosis of Ankylosing Spondylitis, especially during the phase of onset, a knowledge of the roentgenographic image of the normal sacroiliac joint is of prime importance, but the images are not always easy to interpret. In practice it is often difficult to know whether a sacroiliac image is normal or pathologic. For this reason we believe it may be useful to give a brief roentgenologic summary of this articulation which, in France, has been the object of special study by P Coriat (45) [1]

Technic Employed for Standard Roentgenography The diagnostic importance of a good sacroiliac image warrants the

[1] We have personally repeated this study in our report *Les articulations sacro-iliaques dans la spondylarthrite ankylosante* Rev rhum. August, 1950 (80).



Figure 54 Antero-posterior roentgenogram of normal sacroiliac joints in an adult.

taking of a well-centered roentgenogram, because one must not attempt to judge the condition of these joints from roentgenograms centered on the lumbar spine or on the hips, and in which one may also" see the sacroiliacs

The antero-posterior position is usually employed the patient in dorsal decubitus, with the thighs slightly flexed in order to diminish lumbar lordosis film under the back, with the tube centered on a line passing through the antero-superior spines of the ilium, and with a caudo-cranial incidence of 20° A compression bulb, applied to the lower part of the abdominal region helps to expel gas from this region but in order to ensure the absence of gas which might interfere considerably with the interpretation of the image, the patient is advised to take, the evening before the examination, two or three spoonfuls of charcoal, and to evacuate the bowel before the roentgenogram is made. In the majority of patients this incidence also brings out the disc between L-4 and L-5



Figure 55 Antero-posterior roentgenogram of the articulated ilium and sacrum (dry bones) Interposed is a sheet of tin which has been cut along the contour of the articulated surfaces.

Oblique Roentgenograms Although this incidence was first advocated long ago (1937) (216), it has not been adopted in routine practice, because its technic is difficult and the information which it yields is not superior to the information obtained from antero-posterior roentgenograms (on the whole at least) in the search for signs of Ankylosing Spondylitis

Roentgenography of the Normal Sacroiliac Joints Antero-Posterior Roentgenograms of the Sacroiliac Joint in the Adult Male In the living subject the articular space (Fig 54) appears to be composed of three elements the anterior branch, the posterior branch and the foot

The anterior or external branch is formed by the overlapping of the

anterior border of the articular surface of the ilium and of the anterior border of the articular surface of the sacrum It is nearly always visible, generally regular, and only slightly undulating Its upper part is outside the sacroiliac joint. Its *lower extremity* takes part in the foot of the joint The elbow in the anterior branch corresponds to the *anterior border of the joint*

The posterior or internal branch is often less clearly visible, and its upper extremity, as well as the summit of the articulation, are not always apparent. It is formed by overlapping of the posterior border of the articular surface of the iliac bone and of the posterior border of the articular surface of the sacrum. Its *lower part* participates in forming the foot of the joint. Above the foot, it usually bends and always runs frankly outward and at some distance from the posterior inferior spine of the ilium, whence it becomes more or less visible This branch corresponds to the posterior border of the joint.

The *foot of the joint* is formed by the lower extremity of the anterior and posterior branches. It *extends* from the bottom of the interspinous notch above to the lower border of the posterior inferior spine of the ilium below The foot is the only part of the image where the interspace is clearly caught by the incident rays The roentgenographic images of the foot of the joint vary greatly On the one hand, the three component elements of the sacroiliac joint may be *more or less visible* and, on the other hand they may *group themselves in a different manner*, and thus the sacroiliac joint may present different pictures

These variations may be related to

The incidence at which the roentgenogram is made

Individual morphologic variations according to age and according to sex.

In children between *eight and 12 years of age* the sacroiliac joint space is still wide and single The sacral vertebrae fuse one with another and form a more or less regular line. The inner portion of the iliac wing extends inward beyond the border of the first sacral vertebra, and its inner border which is parallel to the border of the second sacral vertebra, may give the appearance of a second joint space comparable with the posterior branch of the articular space in adults (Fig 58)

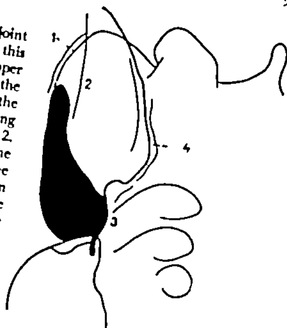


Figure 56 Diagram 1 False joint space above the true joint space this is due to overlapping of the upper extension of the anterior contour of the auricular surface of the ilium and of the upper part of the contour of the wing of the sacrum above the joint space 2 Posterior semi-articular line of the external iliac fossa 3 Foot of the joint. 4 False articular space due, on the one hand to overlapping of the contour of the iliac crest and of the posterior superior spine of the ilium, and on the other hand, of the sacral head formed by the tubercles.

In adolescents when the sacrum nears the end of its development in length, an epiphysis appears at its lateral border in good roentgenograms this epiphysis is visible at the level of the three first sacral vertebrae in the form of a long band (Fig 59a)

The sacroiliac joints of *old persons* (80) have distinct articular spaces a roentgenographic image of ankylosis is altogether exceptional Out of 250 roentgenograms M P Weil and Sichère (217) found a bilateral sacroiliac ankylosis in 2% of the cases, that is, in a statistical proportion without significance, perhaps, in old persons, tomography of the sacroiliac joints might show partial fusion of the joints which are not visible in the standard film—fusions such as those found by Sashin (186) at necropsy

Errors of Interpretation (*Figs 55 and 56*) One should not regard as joint contours

The supra articular part of the anterior joint space

The intra articular and para articular surface lines owing to the complicated shape of these articular surfaces, roentgen rays that are tangential to these asperities' yield roentgenographic images called surface lines, which should be distinguished from the articular contours

The false articular space, which follows the inside contour of the posterior superior spine of the ilium.

The gluteal or posterior semi-circular line

To these causes of error must be added those which have already been mentioned gas and fecal matter in the bowel

TOMOGRAPHY OF THE NORMAL SACROILIAC JOINT *In adults* (from a study made on 5 subjects), the tomographic sections were made in a frontal direction at a distance of *4 to 9 cm* from the posterior plane, and thence *at every centimeter*

In the section made at 3 or 4 cm (Fig 57a), according to the thickness of the subject, one may see very clearly the *contour of the posterior superior iliac spine* (plane of the iliac wing) and the *foot* or posterior part of the articular space

At 5 cm the foot of the joint space extends upward some segments of the posterior border of the joint space are often seen

At 6 cm (plane of the sacrum) (Fig 57b), the entire articular space is visible The entire contour of the sacrum is outlined by a continuous line The first sacral foramina are very clearly visible. The foot, which is vertical and straight, extends upward the upper part of the joint space, which curves like the arc of a circle, branches, like a graft, a little below the upper end of the foot *This section made at 6 cm is the most important one for study of the articular changes*

At 7 cm the entire length of the articular space is visible it is slightly more convex and is farther out than in the previous plane

At 8 cm. the articular space, which remains single continues its outward displacement The foot is still slightly visible

At 9 cm. the articular space is still farther outward

Comments on the Tomograms 1 This description applies to subjects of medium height and weight The position of these different images may vary from 1 to 2 cm according to the thickness of the individual

2 Tomography enlightens one especially on the condition of the articular surfaces that border the joint space. The contour of *this space, which is 2 mm wide, is sharp* especially in the portion called the foot. In the event that the joint space on one side disappears, a pathologic image must not be inferred if in making the roentgenogram, the conditions essential for symmetry of the pelvic bones have not been fulfilled

In adolescents (Fig 59a) the contour is irregular in the section sacral marginal plates corresponding to the epiphyses (Figs 59b and c) may be seen and this is useful for differential diagnosis.



Figures 57a and b Tomography of the normal sacroiliac joints of an adult. a. Section made at 4 cm from the posterior plane plane of the iliac crest the foot of the articulations may be seen very clearly b Section made at 8 cm. from the posterior plane "plane of the sacrum" one may see the foot and branching from it like a graft, the middle and upper portions of the articular space.

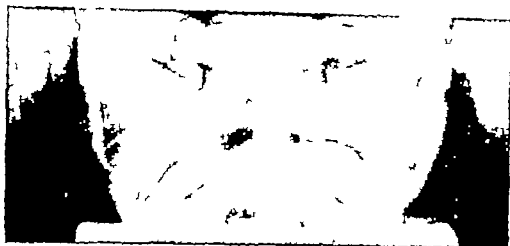


Figure 58 Anteroposterior roentgenogram of the sacroiliac joints of a child 12 years old The sacroiliac joint space is single and very large. Worthy of note is the overlapping of the wing of the ilium and the sacrum.

In old men the sacroiliac joint space shows, in a tomographic section, the same design as in adults, but it appears to be bordered by a longitudinal band of bone sclerosis Often small osteophytic beaks are found at the lower end of the foot (Fig 60)

Tomography permits a better analysis of the sacroiliac changes In cases where in a standard roentgenogram these changes are typical of Ankylosing Spondylitis, such an analysis is not important but in cases where the changes are doubtful or at the borderline of normalcy, tomography may sometimes furnish information which is useful for diagnosis

Roentgenology of the Sacro-iliac Joints in Ankylosing Spondylitis In Ankylosing Spondylitis, *at the present time* we consider *roentgenographic involvement of the sacroiliac joints as a constant* sign As a general rule it appears early and precedes the appearance of the spinal symptoms

While reviewing the roentgenologic case records of more than 200 patients, we again studied the evolution of the roentgenographic image and we again found *the three stages already described by one* of us in 1939 (70)

STAGE I We have usually found this stage in recent cases of Ankylosing Spondylitis in which the disease had progressed clinically from two, three, or six months to five years, and sometimes even in cases where the disease had progressed *for a longer time*

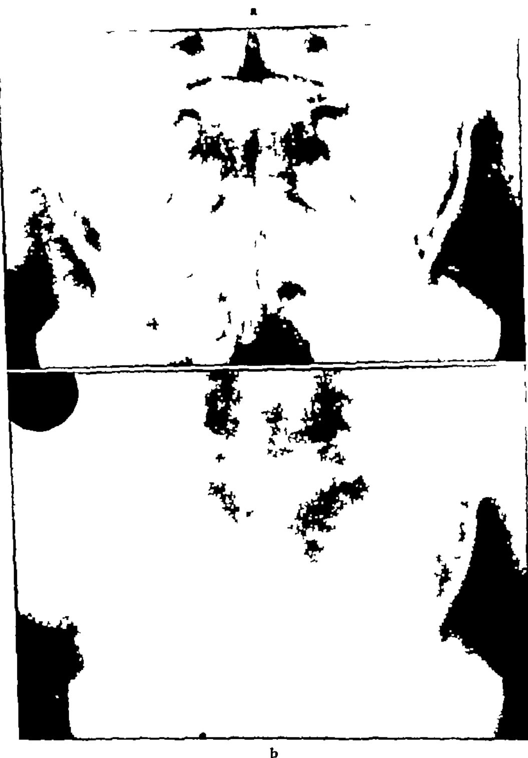


Figure 59 Antero-posterior view of the sacroiliac joints of a young girl aged 18 years a. Standard roentgenogram the joint space is narrower than that of a child the right marginal plate is clearly visible the left plate is less visible. b Tomographic section made at 6 cm. plane of the sacrum" This tomogram shows that the marginal plates, which were visible in Figure 59a were not artefacts.

when the first roentgenogram had been made late We regard it as the first definite stage of the sacroiliac changes at the beginning of the disease

False Widening of the Articular Space In a segment of one or two joints, usually in the lower or middle third, the subchondral zone of the sacral and iliac bones becomes foggy and irregular

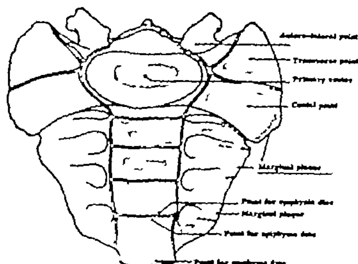


Figure 59c. Diagram of the epiphyseal points of the sacrum, according to Rouvière note the lateral marginal plate.

The sharp borders of the articular space disappear a loss of calcium occurs along the articular border, and this *marginal decalcification* gives the appearance of widening of the articular space (Fig 61)

This widening may be even greater and may thus produce an image similar to that of tuberculosis (Fig 62) A chain of small local widenings may produce the bead form of G Scott (Figs. 63 and 69)

At the same time, and in contrast to the pseudo-widening, there may be found *condensation* in large vertical zones along the entire subchondral region extending especially toward the wing of the ilium (Fig 64)

The degree of pseudo-widening and of bony condensation varies greatly and one or the other may predominate (Figs. 61 and 64) In a few cases only minor changes may be found



Figure 60 Tomogram of the normal sacroiliac joints of a patient aged 60 ye section made at 8 cm. from the posterior plane band of bone sclerosis border the articular space. This should not be interpreted as pathologic.

STAGE II We have observed another roentgenographic ty which, when serial roentgenograms are available, follows stage

The joint space is partly or wholly blotted out, and it is replac by a *change in structure which affects the entire articular zone* as far a and 3 cm from the articular space

The bony condensation is less pronounced and less extensive th at the outset, but it presents a *mottled and striped appearance* (Fig 6

STAGE III We consider this as the terminal stage because have observed it in the latest roentgenograms of the same roentge graphic series

The joint space has completely disappeared in the lower tw thirds of the sacroiliac joint, and sometimes also in the ligament portion bony ankylosis has developed (Fig 66) Sometimes a vestiges of the articular space may be seen

The condensation has disappeared and the bone has recovered normal density or nearly so In some very advanced cases a se of fine bony bands may be seen they follow the lines of force t prolong the column of the lumbar interapophyseal articulati

Figure 61

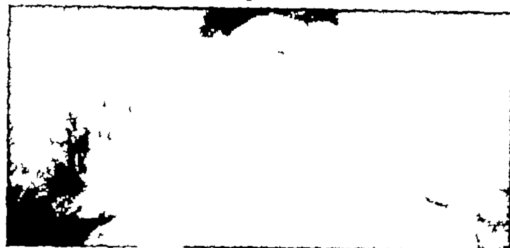


Figure 62

Fig 61 Mr Pif 17 years old with Ankylosing Spondylitis of $2^{1}/_{2}$ years duration bilateral sacroiliac changes in stage I false widening of the articular space and faint peri-articular condensation.

Fig 62. Same patient as in Fig 61 roentgenogram made a month later sacroiliac changes in stage I producing in the film a pseudo-tuberculous appearance "cavity and right sacroiliac sequestrum.

Figure 63

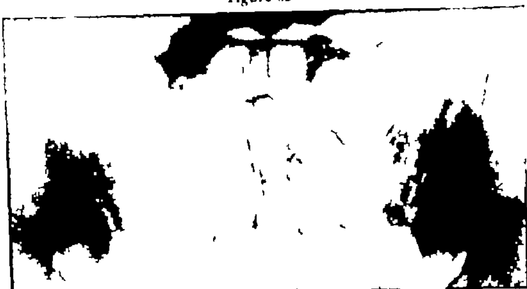




Figure 64

Figure 63 Same patient as in Fig 24 bilateral sacroiliac change in stage I irregular joint spaces with small local widenings producing the "bead" image of Scott.

Figure 64 Mr Dl 29 years old, with Ankylosing Spondylitis of 14 years' duration bilateral sacroiliac change in stage I irregular joint space, but especially marked periarticular condensation.

Figure 65





Figure 66

Figure 65 Mr Au 42 years old, with Ankylosing Spondylitis of 20 years' duration bilateral sacroiliac changes in stage II the hardly visible joint space is covered by a zone of condensation with a mottled appearance.

Figure 66. Mr Alc 51 years old, with Ankylosing Spondylitis of 34 years duration bony ankylosis of the sacroiliac joints.

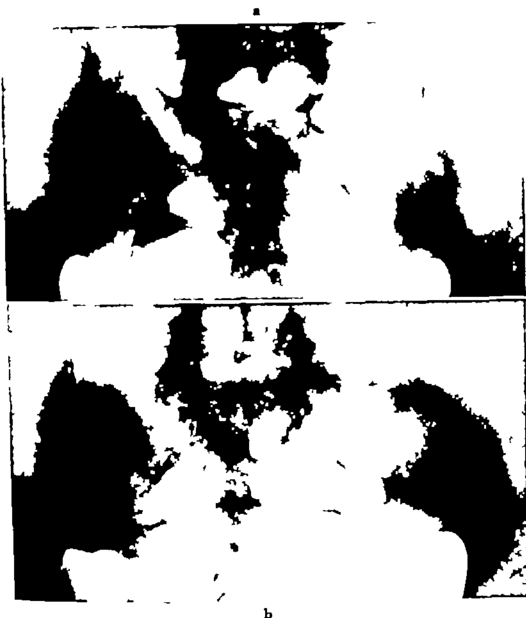


Figure 67a and b. Mr Bur 20 years old with Ankylosing Spondylitis of two years' duration a. Sacroiliac changes in stage I irregularity of the articular contour and subchondral condensation b Five years later picture of bony ankylosis the condensation zones have disappeared

cross the former sacroiliac joint space, and extend downward and outward towards the upper part of the acetabulum (see Fig 27)

In the series of roentgenograms the sequence of these three stages has been very distinct In three of these series the evolution from stage I to stage III took place rapidly within a period of from three to six years (Fig 67a, b), but this usually requires more time.

RELATIONS BETWEEN THE THREE ROENTGENOGRAPHIC STAGES OF SACROILIAC CHANGE AND *The duration of clinical evolution* The relation between the type of roentgenographic image and the period of clinical evolution varies greatly (Table, Fig 68)

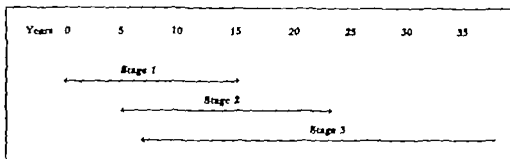


Figure 68 Table showing the periods of evolution of the disease in which the three roentgenographic stages of sacroiliac change (based on 100 cases) are encountered.

Stage I has nearly always been observed during the first months of clinical evolution often during the first few years, but sometimes in rather old cases (the oldest was 15 years)

Stage II has been observed from the fourth year of evolution The oldest case presenting this type of image had been afflicted with the disease for 23 years

Stage III has been observed from the seventh year and has been the only type found in cases of more than 23 years evolution

Therefore, we may conclude that a rough relationship exists between the roentgenographic types and the period of clinical evolution but as the foregoing table shows, for a given period of evolution (from 8 to 15 years) any one of the three types may be observed

Age of the patients Because of the usually slow progress of the disease, the most advanced lesions are observed only after a certain age, but we have not found any particular relationship between the age of the patients and the roentgenographic images

Sex In 50 roentgenographic records of women who were afflicted with spondylitis we have not observed any images that differed from those observed in men

Changes in the Spinal Column Between the stage of the sacroiliac lesions and that of the roentgenographic changes in the spinal column there is a rough relationship

The first stage may be accompanied by syndesmophytes, which are usually not marked, but these are most frequently absent, and they may even be lacking in stage II Stage III is often accompanied by extensive ankylosis of the spine, in a few cases, nevertheless, a definite dissociation exists between the stage of the sacroiliac lesions and that of the roentgenographic signs in the spine, in the resolving forms, for example, we find ankylosis of the sacroiliac joints and a normal spine.

PROBLEMS RAISED BY ROENTGENOGRAPHIC INVOLVEMENT OF THE SACROILIAC JOINTS AT THE TIME OF ONSET (80)

Slight roentgenographic changes—are they reversible? When the roentgenographic images observed in the three stages which have been described are bilateral, they justify a forthright diagnosis of Ankylosing Spondylitis, but at the outset of the disease we have found, especially since we have paid greater attention to its early diagnosis, roentgenographic images of the sacroiliac joints that presented only slight changes. From the point of view of diagnosis, and taking into consideration the intermittent clinical signs during the phase of onset, a study of these images seems to be most valuable

Example Ch Mon , 17 years old July, 1948

Following a 'grippe with pain in the pharynx and fever the patient presented an exacerbation of polyarticular rheumatism in connection with which the diagnosis of acute rheumatic fever or of infectious arthritis was discussed Treatment with salicylates for 20 days, then treatment with Penicillin, did not yield any improvement. Examination discloses 1) An intense torticollis, with the cervical region very stiff and painful 2) the peripheral joints are affected the tarsals, the wrists, and the metacarpal and phalangeal joints of the second and third fingers. The temporo-maxillary joints also are slightly involved

In these joints the spontaneous pain is not severe there is definite swelling, but no redness or local heat. These disturbances are not erratic, but in these articulations they have persisted since the onset. Except in the cervical region, spinal mobility is normal. The respira

tory movements are normal No fever, a good appetite, and no noticeable sweating The heart is normal The rate of sedimentation is 65 mm for the first hour

Considering the features mentioned and the age of the patient, we thought he was suffering from Ankylosing Spondylitis with a peripheral polyarticular onset. The patient was treated with Gold salts with gradual improvement in one and a half months all the symptoms had disappeared.

Because of the complete remission of this polyarticular exacerbation and the patient's age, one might think of rheumatic fever which also presents remissions of this kind On August 31, 1948, a roentgenogram of the sacroiliac joints was made (Fig 69), and this showed bilateral involvement of these joints

Right Sacroiliac fading of the middle third of the articular spaces, and slight change in structure at this level The lower third of the articular space has indefinite borders and seems to be abnormally wide. Similar, but slightly less pronounced, lesions were seen in the image of the left sacroiliac joint

Evolution. May 11, 1949 During the preceding winter, the patient had received treatment with Thorium X (1,100 μg) From the point of view of the spine and joints he had enjoyed a good winter For a month he had suffered from pain in the knees without swelling and some pain in the right side of the thorax, but this pain had not been accentuated by cough On examination objective findings were absent.

A second roentgenogram of the sacroiliac joints (Fig 70), as well as a tomographic examination on December 15 1949 gave negative results.

Summary of this case A patient aged 17 years presented an inflammatory polyarticular disturbance affecting predominantly the small joints of the hands, of the feet, and of the cervical region and continuing steadily for a month and a half and for two years the clinical remission continued Roentgenography of the sacroiliac joints revealed bilateral fading of the joint spaces, and this fading gradually disappeared in the course of one year

This case leads us to consider that

The sacroiliac lesions that are present at the beginning of the disease may be slight and faint, and may consist of simple fading of the articular space, which is masked by a more or less dense and irregular fog

Figure 69





Figure 70

Figure 69 Ch. Mon 17 years old, with Ankylosing Spondylitis with an onset featured by peripheral exacerbations of resolving polyarthritis. Standard roentgenogram of the sacroiliac joints after two months' evolution bilateral blotting out of the joint spaces.

Figure 70 Same patient as in Fig 69 Standard roentgenogram of the sacro-iliac joints 17 months after the onset the joint spaces are again quite visible.

Judging from standard roentgenograms, these early roentgenographic lesions may be reversible[1] and may constitute but a transient outburst, just as the clinical features of this phase may be. If we had seen only the latest roentgenograms of this patient, which showed normal images, we could not have made a diagnosis.

For a long time we have been following a clinically typical case of Ankylosing Spondylitis which has been progressing for 19 years; roentgenograms made nine years ago showed slight sacroiliac changes with widening of the joint spaces (widening probably related to pregnancy), and a subchondral condensation of bone of small extent, but typical syndesmophytes. The evolution of the disease has been benign but typical, and at the present time the disease seems to have become stabilized. A recent standard roentgenogram of the sacroiliac joints, made nine years after the first, shows an image that might be considered within normal limits, but the tomograms reveal changes in the joint spaces (80).

In our study of the different stages of the roentgenographic changes in the sacroiliac joints we have seen that *the bony condensation, which is more or less marked in stage I (stage of onset), gradually disappears during the subsequent stages, leaving a more or less complete bony ankylosis of the joint.* The cases which we have just presented show that, although the early condensation of bone may not be pronounced, it may leave, when it disappears, only a slight alteration of the joint space, and that a normal appearance of standard roentgenograms may make the diagnosis of the disease very difficult. The few cases in which vertebral syndesmophytes and "intact" sacroiliac joints are observed in standard roentgenograms may belong to this group.

Delay in the Appearance of Roentgenologic Lesions. In a few cases, in spite of a clinical picture of spinal involvement which strongly suggests Ankylosing Spondylitis, we have not found any roentgenologic changes of the sacroiliac joints in standard roentgenograms; these cases had progressed for four months, 15 months, four years, and five years, respectively. They raise the problem of delay in the appearance of these changes, a problem which is far from solved.

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[1] *This reversibility of the sacroiliac changes* has been observed in other conditions; one of us (254) brought this out in connection with melitococcic arthritis, and Shipp and Haggart (262) in osteitis condensans ilii.

at the present time A delay of from several months to three years (21) in the appearance of roentgenologic alterations in the sacroiliac joints is admittedly possible but the duration of this latent period cannot be set precisely Moreover, after having noted the possibility of reversal of the sacroiliac changes, we believe that perhaps these cases should be envisaged from this point of view

Isolated Peripheral Involvement at the Outset (78) *and Roentgenologic Lesions of the Sacro-Iliac Joints* Is the appearance of sacroiliac changes concomitant with peripheral articular involvement when the latter precedes spinal involvement by many years? Clinically the sacroiliac syndrome develops early, and it must be regarded as the first site of spinal involvement, but it may be preceded by clinical manifestations in the extremities

In summary, *the ideas of the precocity and constancy of the sacroiliac changes in Ankylosing Spondylitis are always valid in practice* And these changes constitute the most useful sign from a diagnostic point of view As the series of roentgenograms shows, the sacroiliac changes precede the other spinal manifestations

In those rare cases which clinically suggest Ankylosing Spondylitis, but in which changes in the sacroiliac joints are not observed, the finding of typical syndesmophytes enables one to make the diagnosis of Ankylosing Spondylitis In our opinion the slight sacroiliac changes had disappeared without leaving their usual traces, that is, ankylosis But in early cases in which syndesmophytes are absent a diagnosis of Ankylosing Spondylitis should not be made, only subsequent events may enable one to make it.

Thoracic Symptoms

Limitation of respiratory movements and deformity of the thorax, which have occurred with or without pain, are one of the most characteristic and most frequent signs of Ankylosing Spondylitis

Thoracic Pain

We have found this symptom in one fourth of the cases, the pain often supervenes during the first six years of the disease

Three principal seats of pain must be considered

Pain in the Dorsal segment of the spinal column, or posterior thoracic pain, has its origin in the intervertebral articulations, in the costotransverse articulations, or in the costovertebral joints and dorsal muscles, it has already been described in the chapter on dorsal pain (p 59)

Low thoracic, or radiating, pain We have observed that radiating intercostal pain is always low

This kind of pain occurs in bands and most frequently involves several intercostal nerves (D-6, D-7 D-12, and down as far as the abdominal region) This pain is increased by cough. We have seen a case in which the topography of the pain was clearly monoradicular Sometimes the pain is very persistent and, from a diagnostic point of view, it is very characteristic of the disease Usually J Forestier examines the thoracic region by making sudden antero-posterior and latero-lateral pressure (see Examination) pain caused by this procedure may be taken as a sign of progressing disease in the thoraco-dorsal region

High thoracic, sternal and parasternal pain is due to involvement of the manubrio-sternal, chondro-sternal, and chondro-costal articulations

Involvement of the Anterior Thoracic Joints

Chondro-sternal and Chondro-costal Articulations. The swelling is hardly visible and only slightly painful on palpation, and for this reason pain due to involvement of these joints has often led to a wrong diagnosis, when the pain is on the left side, it may be erroneously regarded as cardiac pain and may be attributed to acute rheumatic fever In other cases it has been regarded as a pleuro-pulmonary manifestation

For sometime we have systematically investigated this kind of pain by making direct digital pressure on these articulations, and we have found it rather frequently

Manubrio-sternal Articulations. We have collected three cases, all three in women Here the swelling at the level of Ludwig's angle is quite visible In lateral roentgenograms of the thorax a fusion of this joint with disappearance of the articular cleft, may be seen Savill (259) removed specimens from this superficial joint and studied them histologically (p 282)



Figure 71 Figure 72 Figure 73

Figure 71 Mr Deg 28 years old with Ankylosing Spondylitis of 10 years' duration. Dorsolumbar ankylosis and cervical stiffness. Patient seen from behind Prominent spinous processes, especially in the lumbar region (in the lateral photograph the disappearance of lumbar lordosis is apparent) and marked muscular atrophy

Figure 72. Same patient as in Fig 71 Side view showing antero-posterior flattening of the thorax and loss of tone of the abdominal muscles.

Figure 73 Mr Gai 57 years old, with Ankylosing Spondylitis of three years' duration dorsolumbar ankylosis, and cervical stiffening Side view dorsal kyphosis corrected by hyperlordosis of the cervical spine and by widening of the lower part of the thorax.

Sterno-clavicular Articulations Although involvement of these joints is not included in the statistics of thoracic pain, a study of it may not be amiss The extremely superficial situation of this joint brings its involvement well into view Out of 200 cases it was noted 22 times, or in *11% of them* We have also found it in rheumatoid arthritis, with a frequency of 7% (in 100 cases) It was observed at some stage in the course of the disease, from the first year to the

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most advanced stages, after 23 years and even after 38 years duration

Involvement of this kind consists of swelling of one or both sterno-clavicular joints (it was bilateral in one-fourth of the cases), and is accompanied by the ordinary signs of chronic inflammation puffiness, slight heat, and especially pain which is very distinct on direct pressure, but only slight on mobilization of the shoulder girdle The swelling and pain persist for a long time and nearly always leave a definite deformity which is characterized by a firm thickening

Thoracic Deformities

We have grouped these into two major types which occur in morphologically different individuals

In asthenic persons, whether tall and lanky or merely thin The antero-posterior diameter of the thorax becomes progressively flattened (thorax like an ironing board—Figs. 71 and 72) The ribs assume a more oblique slant which is especially perceptible in the middle part of the thorax The entire sternocostal breast plate is lowered in relation to the dorsal spine the whole anterior chest wall, it would seem, is displaced downward and backward This might explain the forward and downward projection of the shoulders This flattened thorax may become immobile or may preserve a small degree of movement it gives the impression of remaining in the expiratory position, and by contrast the abdominal respiration is readily visible

In compact or obese persons The upper part of the thorax presents the same deformity as that which was previously described (downward and backward displacement of the sternum) but the lower part of the chest bells out over the enlarged belly (pyriform thorax) The lateral profile of the chest produces a curious silhouette it is flattened opposite the sternum with a prominent abdomen below (Fig 73) In this type as in the previous one, the respiratory movements of the abdomen are readily visible

When dorsal kyphosis is pronounced (Occiput-to-wall distance more than 15 cm.) The position of the thoracic cage in relation to the remainder of the body is modified the upper portion of the thorax and spine inclines forward while the lower margin of the ribs is displaced downward and backward and thus



Figure 74



Figure 75

Figure 74 Mr Mic 39 years old with Ankylosing Spondylitis of 10 years duration lateral roentgenogram of the thorax the chest is not flattened the sternum is 14 cm. from the dorsal spine noteworthy is the calcification of the costal cartilages. The costal respiration of this patient was completely absent. Figure 75 Same patient as in Fig 8bis lateral roentgenogram of the chest asthenic type, with flattening of the anterior chest wall, the sternum being 8 cm. from the dorsal spine. In the anterior part of the roentgenogram marked calcification of the chondro-costal junctions is present.

extends to the abdomen, causing to appear, above the umbilicus, a deep, horizontal abdominal fold

Roentgenography confirms the clinical findings and particularly the downward and backward displacement of the sternum The obliquity of the ribs, which is very marked in asthenic individuals, is less so in persons of compact build Thus we find the two main types of thorax one completely flattened (Fig 75) and the other flattened only in its upper part (Fig 74)

Influence of Thoracic Involvement (stiffness or ankylosis, and deformity) on Respiratory Function The respiratory range

(p 16) was measured in 200 cases. In 182 cases, or 91%, it was less than the minimal normal range (6 cm), and it was normal in 18 cases, or 9%.

A slight reduction in respiratory amplitude may be noted at the beginning of the disease, and this gradually increases, during the first year, respiratory amplitude was never less than 3 cm., in advanced cases the respiratory amplitude, as a rule, was more reduced (in cases in which the disease had been active more than 20 years amplitudes greater than 2 cm are very rare)

When the thoracic movements are very limited, the patients have a *purely abdominal respiration* At each inspiration, the upper part of the belly forms a large and abrupt forward bulge, and at each expiration this upper part returns beneath the costal margin These *movements of the abdominal wall make a striking contrast* to the flattening and complete immobility of the thorax

Respiratory Capacity

This respiration seems to be adequate for the patients because, when they are at rest, they have a normal respiratory rhythm, but when they exert themselves, they may become slightly breathless. In practice the problem of obvious dyspnea seldom arises in certain cases this dyspnea has given rise, in the patient as well as in the physician to the idea of a cardiac disorder

With a spirometer we have measured *the vital capacity* (the quantity of air exhaled on full expiration after a forced inspiration) We made this measurement in normal subjects and in 10 subjects of both sexes who were *afflicted with Ankylosing* Spondylitis and who presented a diminished amplitude of thoracic movement (0 to 4 cm.)

In the normal subjects the capacity varied between 3 3 and 5 l. (average, 3 5 l.)

In patients affected by Ankylosing Spondylitis it varied between 1 8 l. and 3 550 l (average, 2.310 l.) *In subjects afflicted with Ankylosing Spondylitis therefore the vital capacity had diminished one third*

Mechanism of Respiratory Compensation The thoracic wall is immobile for this reason, respiratory function must be accomplished through movements of the diaphragm. We have studied these movements fluoroscopically by means of orthodiagrams.

This study was carried out in a few normal individuals and in patients afflicted with Ankylosing Spondylitis

We measured in centimeters, on the orthodiagrams, the gap between the extreme positions of the diaphragm in normal and in forced respiration

IN NORMAL INDIVIDUALS During normal respiration, the distance between the positions of the diaphragm on inspiration and expiration, did not exceed 2 cm , on forced respiration it was about

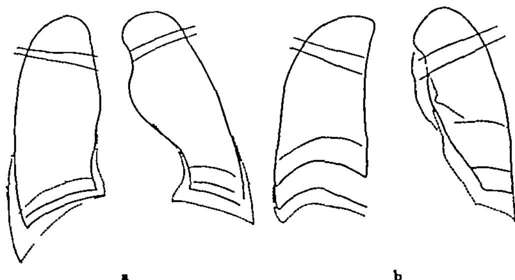


Figure 76 Comparison of the movements of the diaphragm with those of the thoracic wall, during normal and forced respiration in a healthy person and in a patient afflicted with Ankylosing Spondylitis, whose thoracic cage has become fixed. The continuous line indicates the extreme positions of the diaphragm and of the thoracic wall during normal respiration the broken line indicates the extreme positions during forced respiration. a. Normal individual aged 28 years. b Patient 29 years old, with Ankylosing Spondylitis. Here the thoracic wall is immobile the diaphragm, on the contrary shows movements of exceptional amplitude.

4 cm. Because of the separation of the ribs, there was always a notable widening of the lower part of the thorax (Fig 76a)

IN PATIENTS AFFLICTED WITH ANKYLOSING SPONDYLITIS, most of whom presented a marked dorsal kyphosis, the distance between the extreme positions of the diaphragm at normal inspiration and expiration, varied in different cases between 1 5 cm. and 4 0 cm. (average, 2.5 cm) On forced respiration this distance in different cases varied between 5 5 cm. and 11 0 cm. (average, 7 0 cm) Perceptible widening of the thoracic cage did not occur (Fig 76b)

In Ankylosing Spondylitis the amplitude of the diaphragmatic movements



Figure 77 Same patient as in Figure 74 calcification of the lower costal cartilages, seen from the front.

during normal and forced respiration is distinctly increased the cardiac silhouette follows the energetic movements of the left leaf of the diaphragm by moving toward the left side during expiration.

Our observations[1] differ from those of Swaim and Kuhns (210) according to whom the movements of the diaphragm, in patients who are afflicted with Ankylosing Spondylitis, are less pronounced than in normal individuals

[1] Since our study of respiratory function, Hart Bogdanovitch and Nichol (241) have published on this subject a report in which they drew the same conclusions.

Roentgenology We have already described the roentgenographic appearance of the thoracic articulations now we will mention only the *costochondral* calcifications (180) For some time J Rotes-Querol has observed in young patients who are affected by Ankylosing Spondylitis, marked calcification of the costal cartilages

In normal persons the age at which the costal cartilages undergo calcification has been determined by earlier investigations calcification of these cartilages begins at the age of 19 years in the first cartilage (168 169 118) in the other cartilages it occurs later, at ages between 30 and 50 years (168 169) after the age of 50 years, extensive calcification of the cartilages has been found in a third of normal persons Extensive costochondral calcification before the age of 30 is rare however, moderate calcification before the age of 30 years has been observed in 9% of men in women it does not occur before this age.

Köhler (118) observed that, in a few rare cases, complete ossification of all the costal cartilages was found at a very early age this was in persons aged 20 26 34 and 37 years, or even more In all these cases a chronic infection seemed to have been related to these early calcifications but Köhler as well as the other writers who have studied calcification of the costal cartilages, do not mention Ankylosing Spondylitis as a possible cause

We have observed calcification of the lower costal cartilages in six subjects whose respective ages were 21 25 27, 28 29 and 30 years, and who were afflicted with Ankylosing Spondylitis, with a definite reduction of respiratory amplitude

The roentgenographic appearance in antero-posterior and lateral films of the dorsolumbar spine consists of grumous calcium deposits distributed more or less regularly through the lower costal cartilages, and producing a distinct image (Fig 77) We cannot specify the frequency with which this calcification occurs in Ankylosing Spondylitis, because we have not systematically made thoracic roentgenograms of young patients affected by this disease

In four of our cases syndesmophytes were present, but faint, and in two cases syndesmophytes were not present We conclude that, *in cases in which such calcifications occur they may develop early* and may even precede the appearance of the syndesmophytes

From the point of view of pathogenesis calcification of the costal cartilages may depend upon two factors 1) limitation of amplitude of the movements of the thoracic cage 2) the tendency to ossification which in Ankylosing Spondylitis, affects not only the spinal column but also the other articulations

Radiating Pain

In Ankylosing Spondylitis pain very frequently occurs outside of the joints sometimes it is vague, "muscular", and related or not related to static disturbances at other times or in other cases, on the contrary, radiating pain has a sufficiently definite character to permit study and diagnosis

Two main groups of radiating pain may be distinguished 1 Deep referred pain, with a segmental distribution, is due to inflammation of a joint It is of frequent occurrence and, since it is often a manifestation of sacroiliac involvement, it *develops early* 2. Root pain so-called is accompanied by neurologic disturbances paresthesia and superficial sensory disorders, according to the territory of the involved nerve root This kind of pain was studied by Bechterew in 1893 (10) who, as a neurologist, had regarded it as an essential symptom of the disease in our opinion it is less frequent than referred pain and, because it is related to involvement of a spinal segment, it occurs at different periods in the course of the disease. This distinction is especially necessary to interpret pain in the posterior part of the lower extremities, because it is due sometimes to sacroiliac involvement, and sometimes to involvement of the lumbar spine [1]

PAIN WITH RADICULAR DISTRIBUTION—L-5

As far as the lower extremities are concerned, we will deal only with pain of radicular distribution deep referred pain has been studied in the light of the symptoms pain in the posterior part of the thigh, which is caused by sacroiliac disturbances (p 90) pain in the anterior part of the thigh, which is caused by disturbances of the hip (p 93)

[1] *Since our first communication on the symptoms of onset (78), we have been able to determine the precise character of this pain.*

This kind of radiating pain is not of frequent occurrence in our statistics bearing on 200 cases we have noted its presence only 17 times, in all of these the pain had an L-5 topography (example, p 174), we have not observed a single case with an S-1 topography This kind of sciatica was noted at the outset 15 times, of lumbar involvement to which it is related, therefore, this kind of pain is an early sign

The pain was localized in the buttock, in the posterior or postero-external aspect of the thigh, and in the posterior or postero-external aspect of the calf of the leg In two cases it stopped at the heel In another case it involved the external malleolus, and in two other cases it radiated over the dorsum of the foot and of the great toe. The pain may be severe at night but movement often causes it to increase.

In cases in which superficial sensory disturbances have been sought there existed, in two cases, a band like hyperesthesia in the outer aspect of the limb In another case paresthesia with an indefinite localization was present, and in a last case no superficial sensory disturbance was present The patellar and Achilles reflexes were normal Lasègue s sign was found in the 10 cases in which the condition of the joints of the lower extremities permitted the examiner to seek it

The evolution of the pain has a variable duration, sometimes many years and sometimes off and on

In these cases we have not practiced lumbar puncture Ludwig, Short and Bauer have found that the protein content of the cerebrospinal fluid varied between 45 and 121 mg to the liter in 28 6% of a series of 42 cases of Ankylosing Spondylitis This increase in protein, which was most frequently found in very painful or sciatic forms (59), would seem to favor the presence of a "*funiculitis*"[1] due to the neighborhood of the inflamed interapophyseal joints

Elective localization of the pain along the nerve root L-5, which emerges through the foramen L-5 and S-1, seems to be due to the anatomic relations of this foramen, which is especially long and narrow and contains a root which, under normal conditions (30), is larger than the other lumbar roots

When a physician encounters a case of monoradicular sciatica, he thinks practically of compression of the root by a protruded disc

[1] From "funiculus" part of a nerve root that extends between the dural sac and the intervertebral foramen.

We know three cases in which the patients had been operated on *unjustifiably* because the following diagnoses had been made sciatica L-5 due to protrusion of a disc, sciatica favored by spina bifida sciatica at L-5 due to protrusion of a disc associated with Ankylosing Spondylitis In the two first cases a systematic clinical and roentgenographic examination had not been made, and in the third case Ankylosing Spondylitis had not been considered as the possible cause of sciatica At the time of the operation, in spite of exploration at several levels and an extensive laminectomy evidence of a protruded disc had not been found

The treatment is that which is usually employed for pain in Ankylosing Spondylitis here roentgen therapy, with "inflammatory" doses, has one of its most favorable indications

INTERCOSTAL AND ABDOMINAL PAIN

Here a unilateral or bilateral, band like, pain starts in the back and descends obliquely downward and forward towards the median part of the thorax when nerve roots D-6, D-7 and D-8 are affected, and towards the median part of the abdomen when nerve roots D-9, D 10, D-11 and D 12 are affected Our records do not permit us to distinguish, at this level, a root pain from a referred pain.

Sixty six times out of 200 cases, or 55 times at the level of the lower segments D-6 D 7 D-8 and D-9, and 11 times in the abdominal territory corresponding to D-10 D-11, and D-12. We have not found any evidence of a band like pain in the upper part of the thorax.

Early in the evolution of the disease we have found a band like pain 12 times During the progress of the disease, on the contrary we have found this kind of pain 54 times In contrast to pain in the lower extremities, *radiating pain with a band like distribution usually occurs late* and thus corresponds well to the ascending spinal progress of the disease.

In practice this kind of pain is always associated with other symptoms In a few cases the pain may be unusually severe and may give rise to errors in diagnosis pleurisy renal lithiasis (one of our patients had undergone a nephrectomy)

Example of Ankylosing Spondylitis with a girdle pain at the level of D-11 and D 12, which had been attributed to protrusion of an intervertebral disc

J R , 36 years old September 23, 1948 At the age of 27 years, effusion of the left knee with little pain duration, 1$^1/_2$ months At the age of 29 years, without any apparent cause, sudden onset of very severe lumbago which persisted in spite of rest in bed Continuation of the lumbar pain until the date of the record At the age of 30 years, radiating intermittent pain in the buttocks, sometimes in the thighs, and even in the calves of the legs, sometimes on the right side and sometimes on the left

At the age of 35 years, persistence of the lumbar pain, and appearance of a radiating band like pain with a distribution D 11 and D 12, more severe on the left side than on the right A neurologist, thinking that the pain was due to hernia of a disc, had caused an intradural injection of Lipiodol to be made, but this had yielded a negative result. At the present time the patient presents the picture of advanced Ankylosing Spondylitis

Intercostal pain is so characteristic of Ankylosing Spondylitis that its presence should lead the patient s physician to examine him for this disease

PAIN RADIATING TO THE UPPER EXTREMITIES

Of this we have observed six cases at the beginning of the disease and four cases at an advanced stage the pain was concomitant with involvement of the cervical spine Its precise topography has not been determined [1]

[1] In connection with the study of the symptoms of onset (p. 174), a case of probable Ankylosing Spondylitis, which had begun with cervico-brachial neuralgia, was reported (68).

Involvement of Peripheral Joints

We call *peripheral articulations* those of the extremities and those of the scapular and pelvic girdles.

For reasons which have already been mentioned we have classified the sacroiliac joints with the spinal articulations.

Most frequently the patients' functional impairment depends in large measure on peripheral involvement, and especially on involvement of the lower extremities. A second reason that makes peripheral involvement important to study is that too frequently its presence, especially when it develops early, tends to mask the true nature of the disease and to cause errors in diagnosis. We have noted involvement of peripheral joints in three fourths of the cases of Ankylosing Spondylitis: 145 out of 200 cases. Seventy-eight per cent of the female cases, and 56% of the male cases, presented involvement of peripheral joints; in this respect the difference between the sexes, considering the fact that we have only 50 female cases, is not statistically significant.

At the first examination these changes were either discovered in taking the history or they were found by ourselves in two-thirds of the cases (101 cases). At that time they appeared as more or less progressive inflammatory arthritis or the sequelae of previous attacks of arthritis.

The table on p. 129 shows: 1) a predominance of hip involvement; 2) the rarity of "acromelic"[1] involvement. Nevertheless, according to our observations, it does not appear that pure "rhizomelic" involvement is characteristic of the disease: simultaneous involvement of the hips and shoulders is not more frequent than simultaneous involvement of one of these joints and involvement of more distal joints.[2]

[1] Involvement of the distal joints of the extremities, in contrast to rhizomelic involvement of the proximal joints.

[2] Without any doubt involvement of the feet is much more common than we have mentioned, because such involvement often eludes even a methodical examination.

COMPARATIVE TABLE OF PERIPHERAL INVOLVEMENT IN ANKYLOSING SPONDYLITIS AND IN RHEUMATOID ARTHRITIS AT THE FIRST EXAMINATION (FIG 78A AND B)

Joints	*Ankyl Spond in 200 cases*	*Rheumatoid Arthritis in 100 cases*
	%	%
Hips	42 0	10
Knees	28 5	62
Ankles	6 5	58
Feet	11 5	28[1]
Shoulders	30 0	64
Elbows	2 5	46
Wrists	4 5	82
Hands	6 5	94
Temporo-maxillary	6 0	8
Sterno-clavicular	11 0	7

At the time of the first examination these forms of involvement were

Monarticular 21 cases

When monarticular involvement dominates the picture, the diagnosis must distinguish between Ankylosing Spondylitis and an infectious arthritis, and especially tuberculous arthritis

Oligo-articular 75 cases (see Modes of Onset)

Simultaneous involvement of a few peripheral joints, therefore, is very frequent and moreover, usually affects the lower extremities

According to statistics including all the peripheral joints, involvement of the lower extremities occurs twice as frequently as involvement of the upper extremities Therefore, any individual who presents involvement of a few joints, especially in the lower extremities, should undergo a systematic examination of the spine

Polyarticular 1 Involvement with extension approaching that which occurs in rheumatoid arthritis five cases, or 2 5% (2 women and 3 men), therefore, this kind of involvement is rare

At the beginning, or later during exacerbations, of the disease, the peripheral and spinal symptoms may or may not be concomitant. In some cases chronic involvement of the joints of the extremities extended rapidly and, at the time of examination, it dominated the clinical picture Since most of these patients were bedridden the spinal symptoms could be elicited only by a systematic clinical examination and by roentgenograms of the spine

2 Resolving exacerbations of polyarthritis, which are analogous to those of acute rheumatic fever

They are essentially an early symptom 5% of our cases started by an outburst of this type (Modes of Onset, p 178)

ANALYSIS OF ARTICULAR INVOLVEMENT[1]

Involvement of the Hips

In Ankylosing Spondylitis the hip is the peripheral joint most frequently affected (42% of the cases) As far as the percentage of involvement is concerned, there is no appreciable difference between men and women

Involvement of this joint developed in 41 cases before the disease had progressed five years *therefore it occurs early Out of the 17 cases in which it had developed as the first symptom in four cases involvement of the hip remained the only symptom of the disease for many years* (for 5 years, on the average)

These two findings, great frequency and early appearance of hip involvement, are not in accord with what is known of rheumatoid arthritis, in which involvement of the hip is relatively rare (in 100 cases of rheumatoid arthritis involvement of the hip at the time of the first examination which was made after the disease had progressed for varying periods, was noted 10 times moreover, it usually occurs late, we have never observed a case in which rheumatoid arthritis had first begun in the hip) Therefore, there is a striking difference between the two conditions. A chronic inflammatory articular syndrome in which early involvement of the hip occurs should suggest the possible onset of Ankylosing Spondylitis

In three-fourths of our cases this kind of involvement was still present at the first examination Chronic involvement of the hips thus forms a striking contrast to involvement of the knees which in two-thirds of the cases, usually undergoes resolution

Roentgenography makes it possible to classify involvement of the hips into two large groups (F Jacqueline [246]) which seem to correspond to different histologic forms (Pathologic Anatomy, p

[1] The frequency of involvement of a given joint is determined according to the joints found to be involved at the time of the first examination, plus those noted in the history

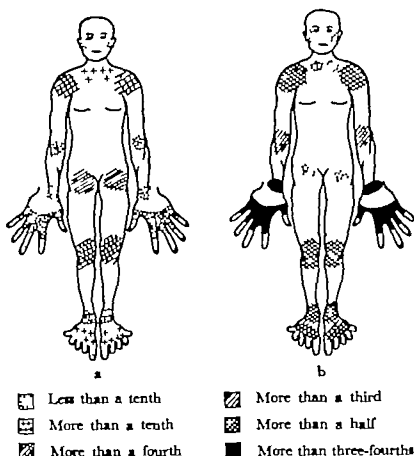


Figure 78 Distribution of Involvement of Peripheral Joints in a, Ankylosing Spondylitis. b Rheumatoid Arthritis. The figures that served as the basis for Ankylosing Spondylitis were obtained from our 200 cases the figures for rheumatoid arthritis were obtained from a series of 100 cases

283) These two anatomic and roentgenologic groups express themselves clinically by symptoms some of which are different.

Ankylosing Form, Without Any Picture of Destruction

This occurs only in young persons, before the age of 40 years It is almost always bilateral. Its onset is often abrupt and its progress is continuous and rapid in less than a few years or even in less than one year the hips may have become fixed The degree of pain varies greatly sometimes it is severe and may force the patient to bed, and sometimes it is not intense, the patient being aware only of a progressive loss of mobility

At the beginning the roentgenographic image is characterized by



Figure 79 Mr Ross 21 years old, with Ankylosing Spondylitis of three years' duration pain in the left hip for one year a. Normal right hip Antero-posterior roentgenograms of the left hip a Picture of slight arthritis osteo-porosis. b. Six months later increased osteoporosis and narrowing of the articular space. c. Five years later bony ankylosis with bony bands, and persistence of the articular space The patient has been bedridden during the period of involvement. d. Seven years after the onset of involvement, an arthroplasty was performed ankylosis recurred rapidly A noteworthy feature was marked bony proliferation at the bottom of the acetabulum



Figure 80 Mr Chau Ankylosing Spondylitis of 15 years duration involvement of the left hip since the age of 12 years picture of bony ankylosis with preservation of the normal width of the articular space

a simple osteoporosis of the entire ilio-femoral region, which may or may not be associated with a slight narrowing of the upper and inner part of the articular space (Fig 79), the contours of the femoral head and of the acetabulum are normal/

Later the picture is one of bony ankylosis the contours of the head and acetabulum are indicated by vestiges which permit us to say that, during the ankylosing process, there had not been any destructive phenomenon Whether the patient is bedridden or not, the joint space may be slightly narrowed (Figs 79c, 80, and 81), most frequently normal, or sometimes widened (246) This roentgenographic space is crossed by fine bony lines which extend toward the head and the acetabular region The osteoporosis may persist, but it is more pronounced in cases of extensive, peripheral arthritis of the spine//

Arthritis with a Picture of Moderate Destruction

//This may supervene either in young persons or, still more frequently, in persons aged between 40 and 50 years The disease may be unilateral or bilateral Pain while the condition is progressing is usually more marked than in the preceding form. The progress of the disease, especially at the outset, is often intermittent. Articular fixation is much less common, and especially less rapid, than in the form with bony ankylosis.

The roentgenographic picture is that of an arthritis which pre

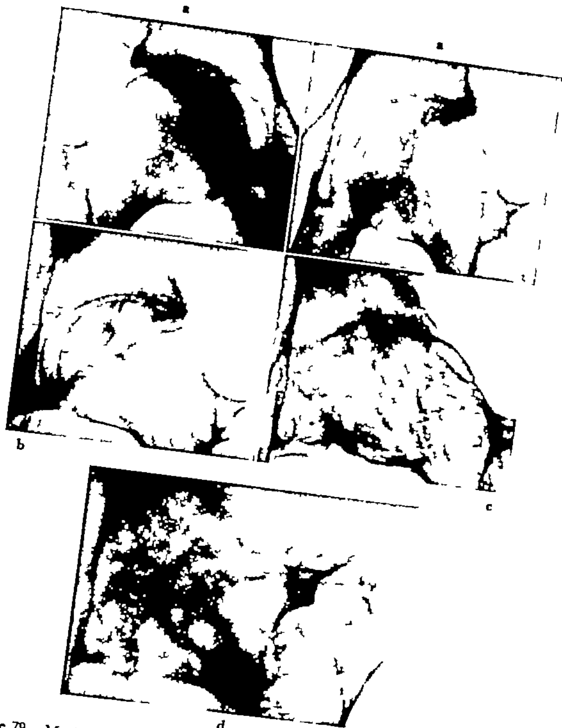


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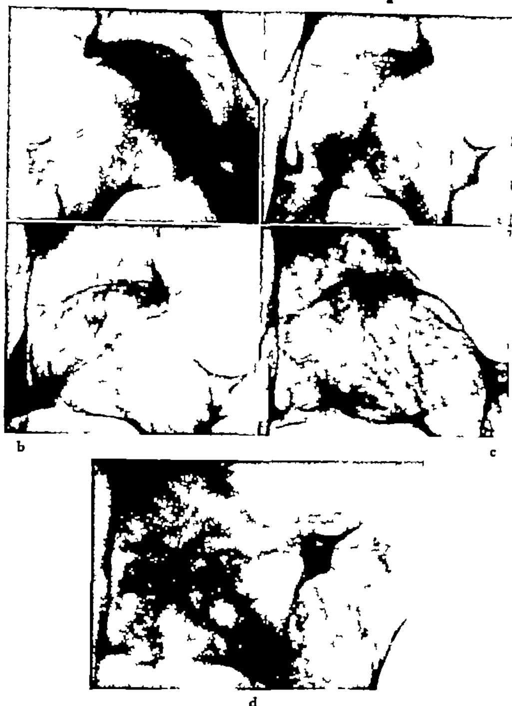


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Figure 81 Same patient as in Fig 83 Roentgenogram of the left hip bony ankylosis, with disappearance of the articular space above the femoral head (the patient had not been bedridden at anytime during this involvement) To be noted is a slight break through of the roof at its upper and inner portion this break through of the roof, which is rare in Ankylosing Spondylitis, is common in Rheumatoid arthritis and is then accompanied by marked lesions of the head





Figure 82 Same patient as in Figure 46 left hip affected for six months. a. Roentgenogram of the left hip with minimal arthritis slight osteoporosis. b Roentgenogram of the right hip with normal structure

sents some peculiarities At the very beginning a simple narrowing of the joint space and slight osteoporosis (Fig 82) this picture must be distinguished from that seen at the outset of other forms of arthritis, and especially from those of infectious character Later there appears a slight irregularity in the contour of the femoral head and especially in that of the acetabulum, particularly in the upper and internal segment of the acetabulum, where even zones of actual disintegration may be present Fig 83 At an advanced stage, the acetabulum is abnormally deep, and its shape has moulded itself to that of the femoral head (Fig 84) but we



Figure 83 Same patient as in Figure 49 hips involved for five years. Antero-posterior roentgenogram of the pelvis. The two hips present typical changes of the crenelated form. Noteworthy are *osteoporosis of the entire region a feathery* appearance of the ischial bones, and irregularity of the surfaces of the pubis.



Figure 84 Miss Per aged 32 years, with Ankylosing Spondylitis of 15 years' duration involvement of the hip for 10 years picture of the crenelated form of arthritis. Note *the spur at the outer part of the head,* indicating a reduction in its volume This form of lysis is encountered especially in the course of rheumatoid arthritis.



Figure 85 Mr Rib 62 years old with Ankylosing Spondylitis of 22 years' duration Involvement of the hips for 18 years. Antero-posterior roentgenogram of the left hip Increased density of the acetabulum, more visible in the upper part osteophytic collar of the femoral head and acetabulum and *narrowing of the joint space*



Figure 90 Mr Vog aged 42 years, with Ankylo Spondylitis of 27 years' du tion markedly progres form. Lateral roentgenogr of the left Knee poste subluxation of the tibial p teau break in the ante part of the plateau, which peared after an attempt straightening the left k shows loss of extension



Figure 91 Same patient as Fig 90 Lateral roentgen gram of the right knee sho the crenelated form of arthri Worthy of note is ossificati of the insertion of the patel tendon.



Figure 92. Mr Gau aged 25 years, with Ankylosing Spondylitis of six years duration Involvement of both knees for two years. Roentgenograms of the right knee. a. Antero-posterior view osteoporosis, erosion of the lower femoral condyle and tibial plateau, and thinning of the internal portion of the articular space. b Lateral view same changes as in the antero-posterior view

prudent walking may prevent stiffening or may permit the patient to recover a satisfactory degree of mobility Here hot spring treatment yields good functional results

As a preventive measure prolonged immobilization of the knees should be avoided by practicing every day active mobilization of these joints, even when the patient's condition requires rest in bed Orthopedic or surgical straightening of defective postures that may affect the hips and spinal column tends to cause the loss of extension of the knees to retrogress.

Proliferative Arthritis Exceptional. The symptoms of this kind of disturbance are analogous to those of the proliferative arthritis incident to rheumatoid arthritis swelling and proliferation of the synovial membrane and this progresses toward stiffening and complete loss of extension Muscular contracture, possibly due to the considerable degree of atrophy seems to play a less important role than in rheumatoid arthritis in a few rare cases,

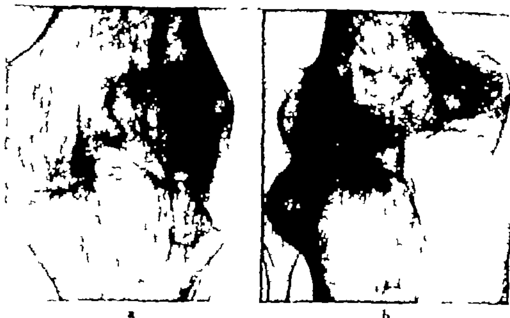


Figure 93 Mrs. Blu aged 40 years, with Ankylosing Spondylitis of 20 years duration panarthritic form, with involvement of the knees since the onset of the disease. Roentgenograms of the left knee. a. Antero-posterior view b Lateral view bony ankylosis with parallel lines of increased condensation giving to the antero-posterior film the appearance of a "curtain" the articular space has preserved a certain thickness. Noteworthy is the ankylosis of the patello-femoral joint.

however, we have observed a posterior subluxation of the tibial plateau (*which is relatively frequent in rheumatoid arthritis*), with or without locking of the patella beneath the femoral condyles In one case this subluxation was accompanied by a break in the anterior part of the tibial plateau (Fig 90)

Usually the articular contours are irregular and marked osteoporosis develops (Figs 91 92a and b) The width of the articular space varies according to whether or not the patient has continued to walk.

Ankylosing Type In very rare cases (see Extensive Peripheral Form) palpation of the hypertrophied and indurated knee gives the impression that the joint is plated with armor the knee is completely fixed the slightest attempts at mobilization make the patient cry out from pain Roentgenography permits one to observe parallel lines of increased density which form a sort of veil which masks the articular space *the thickness of the cartilage may be preserved* (Figs 93a and b)

Infantile type of the Polyhedral Form In two cases in which the knees had become involved at a very early age, that is, at the ages of nine and 12 years, respectively, we have observed a very peculiar form Roentgenograms showed a flattening of the sur faces of the two condyles, these are made up of rectilinear segments that join at an angle, like two blocks of hard and roughly cut stone (Figs 94a and b) The result is that the two surfaces can move freely, but only to the extent of a few degrees, this mechanical limitation is similar to that which is observed in the sequelae of joint fractures To us these deformities of the epiphyseal surfaces seem to be related to the incomplete stage of epiphyseal ossifica tion at the time the knees had become involved

Involvement of the Ankles and Feet

The Ankles

Involvement of the ankles was observed in 6 5% of our cases it had developed early, and in three cases it had been the only symptom of onset of the disease It often undergoes resolution, in half the cases it was still present at the time of the first examination

The severity of the symptoms varies considerably either more or less transient pain and swelling which interfere with walking and which, at the outset, are often regarded as a sprain (these symptoms usually disappear without leaving any sequelae), or more persistent and more severe pain and swelling with the features of a true arthritis perimalleolar swelling and pain on mobilization

The roentgenographic picture shows a simple osteoporosis of the joint or in more severe cases, a distinct osteoporosis and a general narrowing of the joint space (Fig 96) This involvement may be associated with involvement of the foot and may end in more com plex disturbances including a marked degree of impairment

This kind of involvement, especially in cases in which it is isolated, should be distinguished from tuberculous arthritis

Inflammation of the Tendo Achilles

Five of our patients presented tendinitis with pain and swelling, and in one of these cases, the tendinitis was the only symptom of the disease (see Modes of Onset)



Figure 94 Mr Ame aged 20 years, with Ankylosing Spondylitis of 11 years' duration involvement of both knees since the onset of the disease, that is, from the age of nine years. Roentgenogram of the left knee. a. Lateral view b Antero-posterior view flattened and even concave femoral condyles. Image of the knee has a polyhedral form.

The Feet

Involvement of the feet may be localized either in the metatarso-phalangeal joints, with swelling of the anterior part of the foot, pain on walking and gross functional impairment, or in the medio-tarsal region, with swelling of the dorsum of the foot and pain on pressure, or in the region of the calcaneus. In seven cases we have noted pain (spontaneous and on pressure) in the region of the os calcis, or a rebellious pain in the heel. These different kinds of foot involvement are often associated in the same individual.

Involvement of the feet was observed in 15% of the cases.

Eight times it developed at the onset of the disease, and twice it was the only symptom from a practical standpoint, it always occurs early

This involvement may be transient and may subside and dis-



Figure 95 Mrs Clai aged 28 years, with Ankylosing Spondylitis of 16 years duration involvement of the knees for 12 years. Lateral roentgenogram of the left knee polyhedral form. Worthy of note is a complete loss of extension, which was related to a modification of the articular surfaces.

Figure 96 Same patient as in Figure 91 Lateral roentgenogram of the ankle and foot narrowing of the tibio-tarsal joint space, and diffuse osteoporosis of the foot.



Figure 97



Figure 98

Figure 97 Same patient as in Figure 73 Involvement of the feet for three years, with image of hammer toes and toes deviating at an angle, and osteoporosis.

Figure 98 Mr Mer aged 43 years, with Ankylosing Spondylitis of 24 years duration pain in the left heel for two years Lateral roentgenogram of the os calcis of the left foot subcalcaneal and retrocalcaneal bone formations which must be distinguished from ordinary calcaneal spurs.

appear in a few months without sequelae, or it may persist longer and may be followed by stiffness or marked deformity which may be extremely troublesome for the patient toes deviating at an angle, hallus valgus, or hammer toes (Fig 97) In addition to deformities of the toes, flat feet are often found, and frequently this takes the form of external rotation or valgus.

Roentgenography In cases of persistent involvement signs of ordinary arthritis in one joint, or more commonly in several joints, are noted The most characteristic are osteoporosis in the stage of obvious diagnosis, and deformities in the advanced stage Pictures of periostitis (Fig 98) may be noted in the inferior or posterior aspect of the calcaneus Considering the diffuse character of these changes (Fig 96), it is preferable to make systematic roentgenograms of the entire foot

Therapeutic Deductions During the inflammatory period the patient should avoid prolonged standing and walking he should not wear slippers with soft soles that do not provide support for the plantar arch, he should wear shoes or slippers with soles that support the foot and at night the weight of heavy bed coverings over the feet should be avoided by using a hoop and sand bags

In cases of persistent involvement treatment with roentgen rays with careful protection of the toe nails



Figure 99 Figure 100

Figure 99 Mr Del aged 58 years, with Ankylosing Spondylitis of 15 years duration involvement of the left shoulder for two months slight osteoporosis of the head made evident by the pencil line contour

Figure 100 Miss Tam aged 35 years, with Ankylosing Spondylitis of 20 years duration involvement of the right shoulder for 12 years. Antero-posterior roentgenogram showing a very irregular crenelated contour of the head with altered structure. This picture could easily be confused with that of dry caries (Volkman type)

Involvement of the Shoulders

Among our cases the shoulder was affected in 30% Involvement of this joint had developed as the first symptom of the disease only nine times in three of these cases it was the only symptom, and then it had to be distinguished from periarthritis of the shoulder and from certain cases of rheumatoid arthritis with a scapulo-humeral onset.

In slightly less than half of the cases, involvement of the shoulder had made its appearance before the disease had progressed five years, and in slightly more than half of the cases (38 cases out of 60) it was still present at the time of the first examination.

Usually this involvement manifests itself by slight pain around the shoulder, the pain extending to the arm and occurring especially on

movement, and sometimes by marked limitation of movement. On the whole, these functional symptoms do not differ much from those of scapulo-humeral periarthritis. This kind of involvement may end in remission or may persist for many years.

Sometimes involvement of the shoulders develops in cases of severe arthritis of the spine and it may become very intense complete immobilization, marked atrophy of the deltoid, and functional impairment aggravated by a defective posture in internal rotation and adduction which may be very pronounced When the involvement is bilateral, the patient may have difficulty in eating and in attending to his hygienic needs

The roentgenographic picture varies minimal image, slight osteoporosis, and slight elevation of the femoral head in relation to the glenoid cavity (Fig 99), picture of arthritis with erosion of the articular contours, narrowing of the articular space, and osteoporosis (Fig 100), picture of bony ankylosis which is observed especially in the extensive peripheral forms (Fig 101)

Involvement of the Elbows

Involvement of the elbows, which was noted in five cases, was always associated with other symptoms and except in one case, it never occurred at the beginning of the disease

In three cases the shoulder involvement had persisted Limitation of movement, chiefly of extension, was always definite, and in one case the elbow was even ankylosed in a *bad position* In another case marked effusion was present.

Cases with chronic progressive disease show marked roentgenographic changes readily visible osteoporosis of the ulnar epiphysis, irregularly dentate contour of the joints, mainly prevalent in the concavity of the olecranon process Involvement such as this is always associated with other symptoms and does not create any diagnostic problem

Evolution When involvement of the elbows persists, it produces marked functional disability because with an elbow fixed at an angle greater than 90°, the patients can no longer bring food to their mouths or attend to their hygienic needs without help When both joints are immobilized in a position of extension, the



Figure 101 Mr Fre aged 39 years, with Ankylosing Spondylitis of 14 years duration extensive peripheral form with left shoulder involved for two years, and with bony ankylosis (also Fig 110)



Figure 102



Figure 103

Figure 102. Mr Gil aged 40 years, with Ankylosing Spondylitis of 20 years duration severe progressive form. Involvement of the right wrist for three years. Roentgenogram narrowing of the articular spaces, and mottled osteoporosis even when severe involvement is present, phenomena of bone lysis and fusion do not occur

Figure 103 Same patient as in Fig 102. Roentgenogram of the left wrist normal picture

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problem is still more serious, hence the importance of preventing this defective posture

Therapeutic Deductions Place the affected extremity at rest in a plaster splint in a position of flexion at about 80°, which will permit the patient to bring food to his mouth. The application of such a plaster shell, when the elbow first becomes involved, not only calms the inflammatory phenomena, but it also prevents ankylosis in an extended position

When the elbow has already become stiffened in an extended position, forced mobilization under anesthesia may be considered in order to correct the bad position of the affected elbow To enable the patient to recover some mobility of the ankylosed joint an extensive arthroplastic resection is necessary

Involvement of the Wrists and Hands

The Wrists

Only in 4 5% of our cases have we found the wrists affected This is in contrast to the constancy of wrist involvement in rheumatoid arthritis, in one case, however, this involvement, which was then unilateral was the first symptom noted and thus was the only symptom of the disease. In the other cases involvement of the wrist was always bilateral Generally, when the wrists are affected, diffusion of the peripheral lesions is an outstanding feature

Usually the roentgenologic changes are slight, in cases in which they are pronounced (Fig 102) they differ from the changes usually observed in rheumatoid arthritis especially bony fusions, which are so common in the latter disease are never observed in the former But there may be observed bony ankyloses similar to those which are known to occur in the hip joints in cases of ankylosing spondylitis

The clinical picture is that of an ordinary chronic and atrophic arthritis.

The Hands

We have observed 13 cases in which one or several joints in the hands were affected

Usually the disorder consisted of slight and transient inflammatory episodes *in one or several of the small joints in the hand* proximal



Figure 104

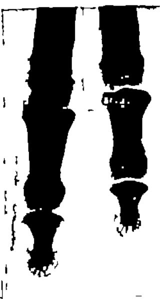


Figure 105

Figure 104 Same patient as in Fig 109 Involvement of the right wrist and hand for 15 years diffuse osteoporosis of the carpal bones especially noteworthy is a lysis of the base of the first metacarpal.

Figure 105 Same patient as in Fig 91 and 96 Involvement of the proximal phalangeal joint of the right middle finger for five months The film shows narrowing of the articular space of this joint swelling of the periarticular soft parts is readily visible.

phalangeal and metacarpo-phalangeal joints *without systematic involvement* (example of hand involvement, Fig 3)

In a few cases there was fibrous thickening of some proximal and distal phalangeal joints, with a more or less marked stiffness which sometimes was associated with pronounced retraction of the flexor tendons. In only two cases, when the disease had reached an advanced stage, the clinical appearance of the hands resembled that of hands affected by rheumatoid arthritis.

Simpson and Stevenson (197), in a study of 200 cases stated that they had not found in any case deformities of the hands and feet similar to those of rheumatoid arthritis In roentgenograms made in 32 cases with clinical involvement they observed, in two cases, diffuse changes consisting of small transparent areas in the diaphyses of the metacarpal and phalangeal bones and these areas were different from areas of erosion found in rheumatoid arthritis and coincided with a general osteoporosis, in 11 cases a simple osteoporosis was present, and in 19 cases anomalies were not disclosed

We have had made only a few roentgenograms of the hands, in one case we found a diffuse osteoporosis, and in another case, a phalangeal roentgenographic change (Fig 105) which, at a relatively early stage, was difficult to distinguish from rheumatoid arthritis.

In Summary, involvement of the hands, besides its rarity does not have the diffuse character which is present in rheumatoid arthritis with its systematic involvement of the proximal phalanges and of the second and third metacarpo-phalangeal joints

Therapeutic Deductions As soon as the slightest tendency to deformity becomes apparent, a half-splint of plaster should be applied in order to maintain a physiologic position.

Involvement of the Temporo-Maxillary Joints

This kind of involvement was found in 6% of our cases.

It was associated with unilateral or bilateral pain in the temporo-maxillary region with moderate swelling and limitation of movement of the mandible

Generally this limitation of movement is moderate and does not raise any therapeutic problem In the extensive peripheral form temporo-maxillary ankylosis often becomes very pronounced and even prevents the patient from opening his mouth In order to feed these severely ill patients, it has been necessary either to extract one or two teeth so as to permit the passage of liquids, or to perform an arthroplastic resection which, unfortunately often ends in recurrence of the ankylosis.

In brief involvement of the joints of the extremities exhibits different localizations than those observed in rheumatoid arthritis, and they may end in bony ankylosis while rheumatoid arthritis causes destruction and sometimes fibrous ankylosis.

Ocular Manifestations

In Ankylosing Spondylitis ocular disturbances (82, 83, 59) are so frequent and so characteristic that they may be regarded as symptoms rather than as true complications

According to different authors, the frequency of these manifestations varies greatly, at a certain period in his rheumatologic practice one of us had the impression that they occurred in from 30% to 40% of the cases, indeed, in our present statistics gathered from 200 cases, these manifestations occurred 17 times, or in 8 5% of the cases.

They usually appear early in the course of the disease, and in three cases they even constituted, in association with pain in the posterior part of the thigh, the entire clinical picture during the early phase of the disease.

CLINICAL FORMS

Classical Form of Diffuse Iritis This will be described first, because it is the most common manifestation

An acute attack, which is rarely bilateral begins by the appearance of perikeratotic injection, often by severe pain extending around the orbit, and by ciliary pain on digital palpation The patient complains of severe photophobia, and sometimes of slight lacrimation, rarely, owing to exudates that form in the anterior chamber vision becomes impaired, most frequently these exudates are fibrinous At times hypopion may occur (Mylius, Streibel, Smeltino)

The attack of iritis may retrogress within from 15 days to a month then the anterior chamber again becomes transparent and, when precautions have been taken, the mobility of the iris is preserved

In these cases the disturbance is a superficial sero-fibrinous iritis which Franceschetti has classified among the typical rheumatic forms by specifying that they may be observed not only in characteristic forms of rheumatism (rheumatoid arthritis and Ankylosing Spondylitis), but also in the course of acute or chronic infectious diseases, or in general or local septic processes (according to him, foci about the teeth, sinuses, or in the intéstine, would seem to be the most common) Usually, when the attack of iritis is short, recurrences, sometimes in the eye first affected and sometimes in the other, are frequent

The most common sequelae of this form of iritis are the synechiae which, because they tend to localize at the margin of the pupil, interfere with its dilatation and assume a semilunar aspect They are often accompanied by a diffuse fogginess of the anterior chamber and by an exudate in the region of the pupil which gives rise to superficial posterior synechiae. After dilatation, these large synechiae give to the pupillary margin an arciform appearance, whereas, in parenchymatous iritis associated with nodular forms (tuberculosis, syphilis), the synechiae are multiple and punctate Thus in Ankylosing Spondylitis, involvement of the uvea manifests itself as a rule by a pure iritis One may observe an endothelial haze, but not gross precipitates, which are symptoms that usually accompany parenchymatous types of iritis (iridocyclitis)

Evidently the sequelae will depend on the more or less severe form of ocular involvement, on repetition of the attacks, and on the treatment employed during the acute phase.

Torpid and Severe Form of Iritis This form, which is more rare, has been observed by Franceschetti and Brocher and by German authors We and P Michaud have observed it in 20% of our cases with ocular involvement in the course of Ankylosing Spondylitis Sometimes it follows recurring attacks of pure iritis, but it may begin from the start with slight manifestations (dull pain and slight disturbances of vision) and may progress insidiously like a severe uveitis that may resemble tuberculous iritis It may be complicated by cataract and by disturbances of the vitreous, and may end in total loss of vision This final result occurred in three of our patients.

Exceptionally progress of the ocular disturbance towards

glaucoma has been observed, it is hardly necessary to recall its gravity

Minimal Iritis Besides the classical form, of which the two modes of evolution have just been described, ophthalmologists admit the existence of a *minimal* iritis Its clinical signs are said to be latent and it is discovered only on systematic examination, preferably with a slit lamp Fombeure and Lacapère (59) have given an excellent description of these forms which apparently manifest themselves only by an insignificant conjunctivitis, without any other functional disturbance than slight fatigue after trying to focus on close objects, especially under artificial light Biomicroscopy, however reveals definite changes in the aqueous and vitreous humors in the form of small particles At a later stage, more important disturbances in the anterior chamber may be associated these are transparent but these forms may leave sequelae that may be recognized only subsequently a granular precipitate and small synechiae in the hyaloid layers

The ocular and articular manifestations do not always coincide, nevertheless, the iritis is more frequent in the severe forms.[1] We have observed iritis in patients in whom, from the articular point of view the disease had become stabilized

DIAGNOSIS

The conditions under which the diagnosis of iritis presents itself may be entirely different according to whether the ocular manifestations are isolated or associated with a clinical syndrome that has few characteristics of Ankylosing Spondylitis

The first possibility will bring the patient to the ophthalmologist who must take into consideration the specific features of the ocular disturbance The superficial sero-fibrinous manifestations should lead him to make a diagnosis of iritis of rheumatic type and to try to find, by questioning and examination, the revelatory signs of Ankylosing Spondylitis. During the period of onset, this diagnosis may be difficult. The inflammatory nature of the iritis may be

[1] Franceschetti and Brocher (84) reported a case in which, in the course of a spinal arthritis, the iritis was associated with phenomena of a non-gonococcic urethritis, thus fulfilling the picture of Reiter's syndrome.

attested by the rate of sedimentation which, during the acute phase, is usually accelerated But when a diagnosis cannot be made from the ocular symptoms, systematic roentgenography of the sacroiliac joints, especially when the patient is young and particularly a young man, should be carried out Likewise, the appearance of rheumatic iritis in a person presenting an indefinitely rheumatic clinical syndrome helps greatly in the diagnosis

When the ocular manifestations supervene during the period of definitely established disease the diagnosis is easier even when the ocular signs are not characteristic, because of the articular, spinal, or peripheral disturbances

PROGNOSIS

The prognosis of iritis in Ankylosing Spondylitis depends on the form of evolution

In spite of recurring episodes of superficial inflammation, most of our patients had only slight impairment of vision, most frequently a few synechiae deforming the pupil, without marked functional disturbances But in spite of careful treatment, a small proportion of our patients suffered a pronounced reduction in the functional capacity of the affected eye, and sometimes complete blindness one of our patients was afflicted with a generalized and ankylosing form of the panarthritic type We do not have any personal records on the pathologic anatomy of iritis in Ankylosing Spondylitis. Some studies of iritis in rheumatic diseases have been carried out but these have not succeeded in specifying the type.

ETIOLOGY AND PATHOLOGY

The etiology and pathogenesis of iritis in the course of Ankylosing Spondylitis leaves much room for uncertainty Most writers who have dealt with this question have classified in the same category *the iritis that occurs in rheumatoid arthritis and that which occurs* in Ankylosing Spondylitis [1] On the clinical plane there is no doubt that it is impossible to distinguish the one from the other but contrary to the opinion that is generally accepted we believe that iritis

[1] In rheumatic fever in principle iritis is never observed.

is much more common in Ankylosing Spondylitis than in rheumatoid arthritis Recently, while reviewing 100 cases of severe rheumatoid arthritis that had been followed more than five years, we found only one case

Since it is now well established that what is called chronic rheumatism is a disease of the mesenchymal tissue, and particularly of the reticulo-endothelial tissue, it is not strange that the iris, which is a mesodermic formation belonging to this type of tissue, should be a reagent for inflammations of collagenous tissue Ophthalmologists know that the iris reacts to all focal infections and that the condition of this organ serves eminently to reveal allergic states [1] No valid explanation of the two following facts has been furnished the predominance of iritis in Ankylosing Spondylitis in comparison with rheumatoid arthritis, and its evolution by acute episodes which are often repeated At one time we had thought that the cervical localization of Ankylosing Spondylitis was an element that favored ocular episodes, but a more careful study of the facts has not shown any relationship

TREATMENT

The first question is the possibility of preventive treatment for iritis. Unfortunately, we are obliged to draw a negative conclusion Moreover, even when Ankylosing Spondylitis is well treated by the general and local methods that have been described elsewhere, an outburst of ocular inflammation may supervene without anything to make one foresee it.

In cases of classical acute exacerbations the physician s first duty is to induce dilatation of the pupil by means of Atropine or Glaucosan, thus protecting the patient from iridian complications caused by the synechiae When the exacerbation is very painful, the retrobulbar injection of 20% Alcohol is indicated During the acute phase and during the ensuing months careful observation by an ophthalmologist is essential in order to track and guard as much as possible against sequelae

[1] In 1938 J Forestier reported a case with flare-ups of iritis at the beginning of each course of treatment with Thorium X, without being able to specify the pathologic mechanism and this observation has been confirmed by other writers.

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Are there any drugs that are capable of radically modifying the progress of an acute attack?

Before replying to this question, it should be recalled that formerly, when rheumatic diseases were not well known, most iritides, the nature of which was not obvious, were regarded as syphilitic and were treated with cyanide of mercury intravenously Happily this practice tends to disappear (Franceschetti recently told us that, in the ophthalmologic clinic of Geneva, he had not seen a case of syphilitic iritis during the past ten years)

Since the attacks of iritis behaved clinically like an acute explosion of allergic type, it was but logical to treat them with shock medication *Intravenous injections of a heterogeneous protein* had given us satisfactory results

Franceschetti and Brocher have used Atophanyl[1] intramuscularly or intravenously The injections are given with a dose of 10 c c of a 10% solution, which are repeated as long as the acute period continues This has given us favorable results

At the present time Franceschetti and his collaborators prefer Irgapyrine (a solution of Amidopyrin in Phenylbutazone) to Atophanyl which is not always well tolerated We employ Butazolidine (Phenylbutazone) in daily intramuscular injections of 5 c c for eight or 10 days

The introduction of Cortisone has added a new drug for the treatment of acute attacks of rheumatic type (Woods, 270) The best results can be obtained at small expense by instilling into the conjunctival sac a standard aqueous suspension of Cortisone acetate The instillation is repeated four six, or even 10 times in 24 hours, according to circumstances. Subconjunctival injection also may be employed The local applications are repeated as long as the inflammation continues. For several days after the inflammation has subsided the treatment should be pursued by the daily application of Cortisone ointment in order to prevent recurrences which often occur when the treatment is stopped too soon The use of Cortisone should not lead one to neglect the simultaneous employment of mydriatics When, during the acute phase, Cortisone is administered intramuscularly or orally it does not have a constant action on the iritis This is also true of A.C.T H

[1] Injectable Cincophen.

Laboratory Examinations

Since Ankylosing Spondylitis belongs to the group of rheumatic inflammations, its diagnosis and treatment profit from the laboratory methods employed for rheumatoid arthritis.

Laboratory examinations may be classified into three groups

Important diagnostic tests

Tests that indicate the activity of the disease and reveal the activity of the inflammatory process

Other biologic and hormonal tests

IMPORTANT DIAGNOSTIC TESTS

Sheep Cell Agglutination Test

The serum of certain patients who are affected by rheumatoid arthritis has the property of agglutinating a larger proportion of erythrocytes in sheep that have been sensitized by a heterogeneous antibody than the normal erythrocytes The anti sheep serum employed is obtained from rabbits that have received repeated injections of sheep erythrocytes

This test has given positive results in from 40% to 60% of cases of severe rheumatoid arthritis (Waaler, Rose, and Ragan, Wager Ball F Jacqueline and A Eyquem)

With modified technics the percentage of positive cases is still higher N Svartz and Schlosmann, by absorbing anti Forsman heterophilic agglutinin, found 90% of positive cases out of 472 cases of rheumatoid arthritis With this technic Van Loghem Langeris found a similar percentage Foz replaced the sheep erythrocytes with Group O human erythrocytes, this makes the test simpler because it is not necessary to absorb the heterophilic agglutinins With this technic he obtains the same percentage of positive cases as N Svartz

This test is practically *negative in Ankylosing Spondylitis* three cases out of 203 serums studied by Ball (223) It is negative in most cases of acute rheumatic fever and psoriasis However, it is positive in certain cases of lupus erythematosus and scleroderma

Streptococcic Agglutination

Cecil, Nichols and Stainsby (226) were the first to show the agglutination of hemolytic streptococci by serum from patients who were affected with rheumatoid arthritis, and since then many papers have been published Boots, Lipman Coss and Ragan (225) found 50% of positive cases in rheumatoid arthritis, and 2% in cases of Ankylosing Spondylitis, but not one positive case in acute rheumatic fever Likewise in cases of rheumatoid arthritis the serum can, in a high percentage of cases, agglutinate suspensions of dead hemolytic streptococcus A (Kalbak [248]) this has not been confirmed in the serum of patients afflicted with Ankylosing Spondylitis

Anti-Streptolysins

The antibodies usually sought for Group A hemolytic streptococcus are anti streptolysin O, anti fibrolysin, and streptococcic anti hyaluronidase These tests are positive in a high percentage of cases of acute rheumatic fever while they are negative in Ankylosing Spondylitis and rheumatoid arthritis.

The different serologic tests are negative in cases of Ankylosing Spondylitis, while they are positive in a large percentage of cases of rheumatoid arthritis and of acute rheumatic fever This point is important, not only from a diagnostic standpoint, but also from the standpoint of classification

Serologic Tests	*Rheumatic Fever*	*Rheumatoid Arthritis*	*Ankylosing Spondylitis*	*Other Joint Diseases*
Agglutination Test on sensitized sheep erythrocytes	0	+	0	0
Streptococcic Agglutination	+	+	0	0
Streptococcic Antibodies	+	0	0	0

Serologic tests in rheumatic diseases. The plus mark indicates a positive result in a high percentage of cases.

ROUTINE EXAMINATIONS

Rate of Sedimentation of the Erythrocytes (E S R)

We employ Westergren's method which can be used by any practitioner

With a syringe of 2 c c capacity and containing 0 4 c c of a 3 8% solution of sodium citrate, 1 6 c c of blood are withdrawn from a vein, this blood is placed in a tube and carefully shaken, this blood is then aspirated into a Westergren pipette (this pipette is a tube with an internal caliber from 2 to 2 5 mm and a length of 30 cm), as indicated by numbered graduations on the tube, the pipette is then placed vertically in a tube holder The rate of sedimentation is read by noting, in millimeters, the height of the clear column of supernatant plasma after one hour, two hours, and 24 hours. We consider as normal figures all those which after one hour, are below 10 mm

Regardless of the stage of the disease, we have found an increased rate of sedimentation in about three fourths of the cases Rather frequently this increase is moderate, and on the average the figures are lower than those obtained in rheumatoid arthritis

Out of 15 cases in which the disease had progressed less than one year 26% had a normal rate of sedimentation

Out of 100 cases in which the disease had progressed more than one year the sedimentation rate was

Normal in	23 cases
between 11 and 30 mm.	47 cases
between 31 and 50 mm.	19 cases
between 51 and 70 mm.	8 cases
between 71 and 90 mm.	3 cases

Out of 40 cases in which the disease had progressed more than 20 years 30% showed a normal rate of sedimentation

The widespread notion that the rate of sedimentation at the beginning of Ankylosing Spondylitis is always abnormally high does not correspond to the truth In our experience, the rate of sedimentation during the first year of the disease is normal in nearly one-fourth of the cases. Later it may increase. Christin (36) ob-

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served six cases in which, during the first year of the disease the sedimentation rate was normal subsequently, in spite of clinical improvement, some of these cases showed an increased rate of sedimentation

On the other hand, our examinations have not confirmed the equally widespread notion that the sedimentation rate returns to normal after the disease has progressed for many years, according to our records, the proportion of normal and increased rates of sedimentation after many years is approximately the same as that found during the course of the disease.

During the exacerbations, the rate of sedimentation increases, but during the remissions it tends to return to the earlier level

Nevertheless, cases are encountered in which, in spite of improvement from treatment, the rate of sedimentation remains abnormally high. In other cases in which, for a long time, the disease has become stabilized a slight increase in sedimentation rate between 10 and 20 mm at the first hour has persisted our experience has shown that these rates of sedimentation, especially when they remain constant, do not indicate continued treatment when they are accompanied by a complete and prolonged disappearance of clinical signs. But with these reservations, the sedimentation rate remains the most useful laboratory test by which to follow a given case

The Blood Proteins

Usually the proportion of total proteins is but slightly changed (260) when a change occurs, it is an increase The albumins diminish, while the α, especially the α 2, and the γ globulins increase (260 232) Fibrin often increases (244) These changes cannot be distinguished qualitatively from those of rheumatoid arthritis, but generally they are less marked than in the latter disease. These changes are also found in cases with signs of clinical progression (232), when they may be observed while the sedimentation rate remains unchanged (244)

Different tests of serum lability Weltman test, gelification with Formol, Cadmium test these often yield positive results and correspond to a change in the globulins (155 101, 244)

The Blood Count

The number of erythrocytes is usually normal, but a slight anemia may be present in 30% of the cases, this anemia seems to be less pronounced than that which occurs in rheumatoid arthritis (155)

The number of leucocytes does not change, apparently leucocytosis does not occur during the active periods of the disease But the differential count may show an increase in the number of lymphocytes at the expense of the polymorphonuclear cells (155)

The significance of blood cytology does not rest on the modifications that may occur in Ankylosing Spondylitis, but on the importance of knowing the condition of the blood cells before and during treatment with roentgen rays, radioactive agents, or gold salts, which may act on the hematopoietic organs

OTHER BIOLOGIC AND HORMONAL TESTS

The 17 Urinary Ketosteroids

Davison, Koets, and Kuzell (50, 231) found that 83% of men who were afflicted with Ankylosing Spondylitis showed an urinary content of 17 ketosteroids to the upper limit of normal, or higher For this group the average was 26 6 mg daily—the ages of the patients varied between 18 and 53 years The low or normal content of other cases may be explained by the clinical condition of the patients and their capacity to respond to stress "A patient with very severe Ankylosing Spondylitis loses his capacity to respond, and his level of excretion is low In cases with benign symptoms the stress is moderate and the level of the 17 ketosteroids is *normal* When the disease has ceased to progress the level of the 17 ketosteroids returns to normal (231)

Of seven men who were affected by rheumatoid arthritis the excretion of 17 ketosteroids had risen in five (the figure ranging between 29 8 and 32.2 mg) but these were young patients who had been ill only a short time and who, moreover were in a phase of clinical improvement

In seven female cases of Ankylosing Spondylitis (women aged be

tween 16 and 40 years) the average excretion was within normal limits, while in women who had long suffered from rheumatoid arthritis the average excretion was always low or at the lower limit of normal.

When these results are compared with those of other writers, distinct differences are evident. These may be due partly to the technics employed and partly to spontaneous variations In order to eliminate this last cause of error it would be necessary to carry out tests over a minimal period of one week. At the present time it would be difficult to draw any definite conclusions

Inorganic Bodies of the Plasma Calcium and Phosphorus

Influenced by the theory of the parathyroid origin of Ankylosing Spondylitis, we had formerly ordered many analyses of blood calcium and phosphorus, but these had always given normal results

The Alkaline Phosphatases

These were studied by Buckley in 1935 (32) Françon and Pellas (87) the acid phosphatases were studied in 1945 by Race (161), and in 1948 by Desmarais (52) These analyses do not permit one to draw any conclusions

On these meager data which, since then have not been confirmed even by their authors, a theory of the pathogenesis of the disease had been advanced (18)

Uric Acid in the Blood

Its level was investigated in a certain number of cases it was practically always normal

Basal Metabolism

In all the cases in which this metabolism was determined it always proved to be within normal limits

The Cerebro-Spinal Fluid

Lumbar punctures have not been done in our patients. Ludwig, Short and Bauer (141) made lumbar punctures in 101 cases of inflammatory rheumatism, in 59 cases of rheumatoid arthritis, and in 42 cases of Ankylosing Spondylitis. The only modifications observed were an increase in the protein content of the cerebro-spinal fluid; this varied between 45 and 121 mg. per liter in 28.6% of the cases of Ankylosing Spondylitis, while it proved to be abnormally high in only 6% or 8% of the cases of rheumatoid arthritis. These results have been confirmed by more recent studies (21), which prove that an abnormally high protein content was present in the cerebro-spinal fluid of 42% of the patients who were afflicted with Ankylosing Spondylitis.

The Synovial Fluids

See Differential Diagnosis (p. 227) and Analysis of Symptoms (p. 140).

Modes of Onset

Early diagnosis of the disease definitely increases the therapeutic possibilities, because defective postures and spinal and peripheral ankyloses, which are the causes of disability, have not yet become established hence the importance of a study of the first symptoms.

The phase of onset is that during which there is not yet any definite and permanent limitation in the mobility of the spine (General Considerations, p 12) It is during this phase that the roentgenographic changes in the sacroiliac joints make their appearance

Time of Onset of the Disease Different writers do not agree in defining the symptoms that constitute the beginning of the disease Most of them regard as the onset of the disease the first spinal manifestations, because they fail to recognize involvement of peripheral joints that may have preceded them or because they consider these as too vague or lacking in objective character

The study of early peripheral involvement, which we have made (78) has permitted us to describe their features and to assert that they have objective characters and thus belong to the picture of the disease Therefore, *we have regarded as the onset of the disease the appearance of the first articular symptoms wherever they may be situated and whether spinal or peripheral*

What has struck us most vividly has been the *diversity of the initial symptoms* which gives to the onset of the disease a disconcerting polymorphism Thus, for example a clinical symptom such as a monoradicular sciatica, a lumbago an iritis, hydrarthrosis of the knee or an arthritis of the hip may by itself and for a rather long time constitute the entire clinical picture at the beginning of the disease

Failure to recognize this period, which antedates clinical involvement of the spine, explains why these patients are allowed to drag along for six or seven years, or even longer without any definite diagnosis and with inappropriate treatment

During this long early period *different symptoms may succeed one another or may combine* to paint the most diverse pictures For example, pain in the posterior part of the thigh and iritis, sciatica and arthritis of the knee

Most frequently the symptoms progress in spurts, *sometimes interrupted by very long remissions* A study of the cases which we have followed for a long time permits us to state that these different spurts are really those of one and the same disease

The phase of onset *may continue for many years* Some idea of this long duration may be furnished by the fact that, in cases in which the onset was featured by peripheral or radiating pain, lumbar involvement made its appearance between six months and 30 years after the onset of the first symptoms (on the average, from six to seven years)

DESCRIPTION OF THE FIRST SYMPTOMS OF THE DISEASE

The first symptoms have been classified for study into three groups according to their topographic situation

Spinal involvement
Radiating pain
Peripheral involvement
and also according to the number of parts involved

Onset by Involvement of a Single Group of Joints

In two-thirds of the cases (131 cases out of the 200 cases in our statistics) the disease begins *with symptoms belonging to one of the three groups previously indicated* For a certain period these "isolated" symptoms may constitute the entire clinical picture

Modes of Onset in Ankylosing Spondylitis

(*statistics based on 200 cases*)

With symptoms belonging to only one of the following groups	131 cases.
Spinal symptoms	82 cases
Radiating pain	7 cases
Peripheral involvement	42 cases
With more complex clinical pictures 69 cases.	

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With more complex clinical pictures	69 cases

These clinical pictures include symptoms belonging to different groups, for example spinal symptoms and radiating pain spinal symptoms and peripheral involvement involvement of peripheral joints and radiating pain.

Onset with Isolated Spinal Involvement

By some writers, even at the present time, spinal involvement is regarded as representing the onset of the disease and, because of this, it should be constant But we have considered it as the "first symptom" of the disease only when it had not been preceded by any symptom of peripheral involvement

We have *observed it in nearly half of the cases* (41% of our cases had an onset characterized only by isolated signs of spinal involvement) *Most frequently the symptoms are confined to a single segment of the spine* sacroiliac, lumbar, dorsal, or sometimes cervical, and sometimes two segments are affected, they are rarely diffuse.

Onset with Sacroiliac Pain In 17% of the cases this sacroiliac syndrome (Analysis of Symptoms, p 85), by itself represented the entire clinical picture at the beginning of the disease The fact that this involvement is difficult to recognize from the case history leads us to think that these figures are below the reality and that sacroiliac involvement will be found more often when it becomes better known and when more patients are observed during the early phase of the disease

EXAMPLE OF THE ONSET OF ANKYLOSING SPONDYLITIS WITH AN ISOLATED SACRO-ILIAC SYNDROME. Sister G aged 35 years. May 21 1941 Six months before, *pain in the buttock and in the posterior part of the right thigh*, extending as far as the popliteal fold. The onset was insidious At the beginning she suffered more when walking or in bed than when standing Four months before, *right sided limp* Since then, she has been in bed half the time, without fever but with loss of appetite

On examination, she was thin, pale, and slender Standing "hippy" attitude toward the right side The spine was supple In the recumbent position, *a point of sharp pain over the right sacroiliac joint* was noted a pain elicited by hyperextension of the sacroiliac joint, and the right hip was free. The sedimentation rate was 12 millimeters A clinical diagnosis of right sacroiliac syndrome was made, without specifying its origin Roentgenograms of the sacroiliac joints showing, on both sides, ill-defined contours and partial disappearance of

the articular spaces permitted us to make the diagnosis of Ankylosing Spondylitis in the phase of onset

Evolution —Patient seen 11 months later extension of pain to the dorsal spine but improvement of the sacroiliac pain On examination, painful points over S-1 and D 5 persistence of the painful point over the right sacroiliac.

September 7, 1942, that is, four months later after two courses of treatment with Radon,[1] abatement of the pain, resumption of activity, and the limp disappeared

During the ensuing years, the Ankylosing Spondylitis progressed in bouts with an increased sedimentation rate.

When the patient was last seen, October 13, 1949, she presented a diffuse involvement of the entire spine But definite stiffness was present only in the lumbar segment a slight kyphosis beginning at D-8 was noted She suffers little, her activity is normal, and the rate of sedimentation is less than 10 millimeters.

In brief this is an Ankylosing Spondylitis that had started with a right sacroiliac syndrome with extension to the spine the following year The disease has progressed in slight bursts, and at the present time the patient's condition is stationary

Discussion The clinical sacroiliac syndrome, clinically unilateral with limping coinciding with a roentgenographic image showing bilateral sacroiliac changes, made the diagnosis possible Extension to the spine, after the disease had progressed one year, and the successive episodes have confirmed the diagnosis.

EXAMPLE OF ONSET WITH PAIN REFERRED TO THE BUTTOCK AND TO THE POSTERIOR PART OF THE THIGH Mr B , aged 26 years September 1, 1949 At the age of 17 years, violent pain in the right buttock and in the posterior part of the right thigh, more severe when the patient was lying down This pain lasted three months Then from time to time pain developed in the posterior part of the left thigh, and then again in the right thigh, and the pain continued to shift from one side to the other One year later, lumbar pain in the form of painful stiffness on awakening in the morning Progress in short spurts, roentgenography disclosed bilateral lesions in the sacroiliac joints (stage I) which permitted us to make the diagnosis of Ankylosing Spondylitis.

Lumbar Onset. This is the mode of onset that is considered classical but, as an isolated symptom, it is *much less common than it*

[1] Radium emanation.

have observed this kind of pain as the first isolated symptom of the disease usually it is associated with lumbago

EXAMPLE OF MONORADICULAR SCIATICA AT L-5 AT THE BEGINNING OF ANKYLOSING SPONDYLITIS. Mr Ver aged 25 years July, 1948 At the age of 23 years, alternating right and left sciatica lasting one year Then, after a six months' remission, recurrence of sciatica with the same features as before, but accompanied by lumbar pain and stiffness, which gradually increased

On examination, the sciatic features were as follows spontaneous pain occurring in short bouts two or three times a day, particularly at night walking caused the pain to diminish, but it was accentuated by cough and sneezing The distribution of the pain was definite buttock, calf of the leg dorsum of the foot, and great toe (15) Distinct band of hypoesthesia, with the same distribution as the pain.

This pain of radicular origin should be distinguished from pain referred to the posterior part of the thigh The latter is frequent and should lead one to think of sacroiliac involvement which, for us, is the cause of it (example, onset p 85 and 170)

Thoracic or Abdominal Girdle Pain Although thoracic or abdominal girdle pain occurs frequently in the course of the disease, it rarely occurs at the beginning and in only two cases was it the only symptom of onset. *In practice, the determination of the origin* of intercostal pain will be helped by analysis of the associated symptoms

Pain Radiating to the Upper Extremities This pain is rare and nearly always it occurs late in the course of the disease. Nevertheless, we believe we should mention one case in which Ankylosing Spondylitis seemed to have started with a cervicobrachial neuralgia.

EXAMPLE OF ANKYLOSING SPONDYLITIS WHICH SEEMED TO *HAVE STARTED CLINICALLY WITH A CERVICOBRACHIAL NEURALGIA* Mr S aged 50 years. July 6 1949 At the age of 48 years, right cervicobrachial neuralgia in the distribution of C-8 cervical region shoulder outer aspect of the arm and forearm and fourth finger The pain disappeared after eight sessions of roentgen treatment to the neck. At the age of 49 years, recurrence of cervicobrachial neuralgia which, this time, was not improved by roentgen treatment A plaster cast to support the head and neck, and to maintain its position relieved the patient A month later, when the cast was removed, the neck remained stiff

At the present time the patient has some pain in the cervical region and in the territory of C-8 but the pain is much less severe On examination, the head is inclined slightly forward and toward the left side Cervical mobility is greatly reduced, extension 4 cm,[1] flexion 2 cm right rotation 10° left rotation 10° right lateral inclination none left inclination 5° pain points C-5 and C-6 Besides the foregoing, we found lumbar stiffness and pain points over the spinous processes of D 10 and D 11, and a distinct reduction in respiratory expansion (2.5 cm) The rate of sedimentation is 14 mm at the first hour Roentgenograms show a faint beginning of syndesmophytes at the level of C-4 and C-5, typical syndesmophytes between D 11 and D 12 and between D-12 and L-1 The sacroiliac joints do not present any visible lesions In spite of the absence of roentgenographic involvement of the sacroiliacs in the standard films, we have made a diagnosis of Ankylosing Spondylitis for the following reasons cervical stiffness much more pronounced than that observed in osteoarthritis, lumbar stiffness, limitation of respiratory expansion, increase in rate of sedimentation, and typical syndesmophytes in dorsolumbar region

Onset with Peripheral Involvement

It is interesting to read over Pierre Marie's first case (143)

Case 1 Harr Louis, aged 31 years, fruit and vegetable merchant, admitted to Bicêtre Hospital February 24, 1896, had been well until the age of 12 years at this moment, *for the first time*, the patient suffered from *pain in the knees*, and the pain slowly progressed. This attack never required him to rest in bed, and the joints were neither swollen nor red the patient did not feel any crepitation, but he had a sensation of violent cramps until the age of 18 years, the symptoms did not progress further the patient was like everyone else, and he did not present any deformity without any difficulty he could cultivate his land

At the age of 18 years, there supervened a *slightly painful* and moderate swelling of the *left knee* and at the same time a very violent, lancinating pain *in the left hip* developed For a few days the patient walked with difficulty, with sticks, and he could make little use of his left leg for four months, after the return of the painful episodes in the left leg our man was condemned to relative rest, but he did not have any fever the lancinating pain remained the only symptom

[1] Distance from chin to manubrium.

'At this moment, *the two knees* but especially the left knee, seem to have been the seat of *a slight effusion* These incidents persisted until August, 1883

'In December 1883 because of a severe coccygeal pain which forced him to lie on his belly until August, 1884 the patient could no longer remain either in a sitting or standing position During this time, the pain in the left leg persisted, but it was less severe than before because of his prolonged immobilization, and after the right hip had become progressively affected it became ankylosed since the month of September 1883 On August 5 1884 the coccygeal pain having diminished, the patient began to get up then a period of abatement supervened until December, 1885

For some time the patient was able to get along without too much difficulty and without much pain But after an interval of one year in December 1886, the vertebral column underwent progressive stiffening lost its suppleness, and finally became ankylosed at this moment, also the movements of mastication became very painful (muscular pain?) In December 1886 therefore, the knee joints were also intact, but the two hips and the spinal column were ankylosed. The cervical region was the last to be affected.

Still, while Pierre Marie described the principal features of the disease, he remembered only the spinal and radicular involvement and thought that the phenomena which he had observed in the peripheral joints were only algias without objective signs, which contradicted his remarkable case This notion that the peripheral lesions at the outset consisted only of vague algias persisted for many years, and the existence of a peripheral phase, although quite *characteristic and sometimes preceding vertebral involvement*, was not recognized

In 1936 Campbell Golding (35), from his observation of a series of 127 cases, affirmed the existence of an onset with involvement of peripheral joints before the appearance of spinal symptoms, and he showed that this mode of onset is even more frequent than onset with spinal involvement. For him this peripheral involvement consists either of vague symptoms of fibrositic type, of polyarticular episodes similar to those of acute rheumatic fever, or of involvement of one or of a few joints, with objective symptoms of arthritis.

In 1942, Gilbert Scott (192) insisted on the existence of a prespondylitic phase, in which the diagnosis can be made only by the

roentgenographic changes in the sacroiliac joints, and he also insisted on the importance of treatment during this phase

At the present time different writers do not agree on the importance and significance of these peripheral symptoms of the disease Quite recently Boland (22) wrote "In approximately one out of 10 patients a history of migratory peripheral joint muscle complaints, with or without stiffness, may be elicited Such complaints usually are not impressive They often have characteristics which suggest an intramuscular and periarticular "fibrositis' and more correctly should be considered as a manifestation secondary to rheumatoid arthritis of the spine With the exception of those patients having a preceding rheumatoid arthritis involving peripheral joints, the first definite rheumatic complaints are almost always located in the back."

Our studies on the symptoms of onset of the disease (78) permit us *to affirm the existence and importance of peripheral involvement during the phase of onset* In 21% of our cases involvement of peripheral joints preceded all spinal symptoms or radiating pain In these patients the time that elapsed between the onset of peripheral involvement and that of spinal involvement varied between six months and 21 years (average, 7 years) We feel completely justified in asserting, therefore, that there is a clinical peripheral phase of the disease, the duration of which varies and which may sometimes continue for many years

The figures which we have cited are probably less than they should be, because, since we have questioned patients more closely in preparing our more recent records, we have elicited this peripheral onset much more frequently than before it is possible that more than a third of the cases begin with peripheral clinical manifestations

These may consist of

Transient episodes of polyarticular arthritis.

A polyarticular syndrome analogous to rheumatoid arthritis (this is exceptional)

Arthritis of a few joints, or even of a single joint.

An unilateral or bilateral inflammation of the Achilles tendon

Algias.

Iritis

Onset with a Transient Polyarticular Outburst. Five per cent of our cases (10 cases out of 200) had begun in this manner

From a practical point of view, this resolving polyarticular involvement appears only at the beginning of the disease, and it often precedes the spinal symptoms

These episodes last a few months (from 2 to 6 months) but in certain cases involvement of one or several peripheral joints may persist for a longer time

These outbursts of peripheral joint involvement may be repeated at widely separated intervals before the spinal involvement develops

Therapeutic medication diminishes the duration of these bouts. Gold salts are indicated, and often from the very first injections clinical improvement occurs In these cases Sodium Salicylate is not as effective as it is generally known to be in acute rheumatic fever but it may be used as an analgesic

Three of our cases of Ankylosing Spondylitis had begun with polyarticular attacks identical to those of acute rheumatic fever, and associated with cardiac signs One of the cases even presented a chorea (Interpretation of these associations, p 286)

Onset with Polyarticular Syndrome (Chronic from the Beginning) Peripheral symmetrical, acromelic and persistent polyarticular involvement, similar to that of rheumatoid arthritis, is rarely discovered in the phase of obvious diagnosis, and still more rarely in the phase of onset persistent involvement of the phalangeal and metacarpo-phalangeal joints as a sign of onset of the disease is exceptional

Onset with Involvement of One Joint or of Few Joints In this group we have gathered the cases in which five or less joints were affected The separation between this group and the previous ones is rather arbitrary It should be noted that the distribution of involvement is sporadic and not symmetrical and that these oligo-arthritides or monarticular arthritides are often chronic.

In 14% of our cases we have found them as the *first symptom and as the entire clinical picture* of the disease.

Example of Peripheral, Oligo-Articular, Onset Mr Ga aged 38 years. Butcher July 19 1949 At the age of

20 years, painful swelling of the knees that confined him to bed for a month

At the age of 25 years, recurrence, but with less severe pain and swelling

At the age of 31 years, swelling and pain in the left ankle, then in the knees, and the pain continued for four years

At the age of 35 years, lumbar pain, followed by rapid extension to the entire spine

Now, the patient having reached the age of 35 years, advanced Ankylosing Spondylitis, with typical involvement of the spine and persistent involvement of the knees, with loss of extension of the left ankle both hips, and both elbows

Although these arthritides may affect several joints from the beginning, the articular inflammation may often begin in a single joint, producing a *picture of monarticular arthritis* for many months, or even for years. Later other joints become involved, and we are then in the presence of an oligo-arthritis In other cases the picture may be oligo-articular from the start, but since the involved joints, save one are affected by a resolving inflammation, a monarticular picture may thus be obtained Finally, we have observed in some cases the alternating involvement of two symmetrical joints, producing what might be called successive monarticular pictures

Thus, considering these different modes of evolution, the diag nostic problem may be to distinguish between a true monarticular disease when involvement of other joints has not been present, and a pseudo-monarthritis in which a carefully recorded history or a systematic examination would reveal multiple joint involvement. This monarticular inflammation should be differentiated from tuberculous monarthritis (Diagnostic Problems, p 225)

EXAMPLE OF A MONARTICULAR ARTHRITIC ONSET Mr Ro , aged 27 years September 21, 1937 At the age of 18 years, right sero-fibrinous pleurisy

Three months later appearance of left pulmonary lesions (T B ?), which were treated with oleothorax

At the age of 20 years, bilateral pulmonary lesions and at the same time marked effusion of the left knee with fever this persisted for two years—patient confined to bed. Guinea pig inoculation negative.

At the age of 23 years, involvement of the right knee These arthropathies continued until the age of 24 years when, after immo-

bilization in a plaster cast, both knees became ankylosed in an extended position At this time the pulmonary lesions were stabilized

When the patient got up, he realized that he also had an ankylosis of both hips and of the spine he thus presented the typical picture of Ankylosing Spondylitis

In brief, the patient was affected by an Ankylosing Spondylitis which had started with effusion of the left knee that persisted for two years and was the only clinical symptom

TABLE OF UNILATERAL AND BILATERAL (SYMMETRICAL) INVOLVEMENT AT THE BEGINNING OF THE DISEASE

(*present as an isolated symptom in 21 out of 200 cases*)

Hip	4 cases
Knee	8 cases
Ankle	3 cases
Foot	2 cases
Shoulder	3 cases
Elbow	0 case
Wrist	1 case
Hand	0 case

Onset with Inflammation of the Achilles Tendon This form of onset as an isolated symptom is rare we have observed it but once [1]

EXAMPLE OF ONSET WITH INFLAMMATION OF AN ACHILLES TENDON Mr Ma physician aged 26 years August 3, 1948 At the age of 20 years, progressive inflammation of an Achilles tendon with very severe spontaneous pain.

On examination the tendon was slightly swollen red, and slightly painful Treatment with roentgen rays caused the tendinitis to retrogress

During the ensuing months, appearance of a sensation of spinal stiffness and high dorsal pain

Now, after six years of evolution, the patient presents a typical Ankylosing Spondylitis, with stiffening of the entire spine

Onset with Algias Formerly and even today, for many writers, vague algias represented the entire pre-spondylitic phase. A more careful analysis of the symptoms of onset has permitted us to verify the fact that, in most cases these are definite objective manifesta

[1] Since the publication of the French edition, we have seen two other cases.

tions related to articular involvement Nevertheless, a few cases remain in which the onset of the disease consisted of more or less vague algic manifestations

EXAMPLE OF ONSET WITH ALGIAS Mr So , aged 58 years June 7, 1939 At the age of 38 years, articular pain in the foot, knee, and shoulder, without any swelling Since then, erratic, intermittent pain

At the age of 55 years, appearance of lumbar pain, and the patient tends to become round-shouldered Now, on examination, the patient presents a typical Ankylosing Spondylitis

Onset with Iritis This was observed in only 1 5% of our cases, and once the iritis was the only symptom at the beginning of the disease

EXAMPLE OF ONSET WITH IRITIS Mr Ro , aged 55 years. August 13, 1947 At the age of 25 years, attack of iritis in the right eye multiple recurrences alternating in one eye after the other until now At the age of 39 years, pain radiating to the posterior part of the lower extremities and alternating from one side to the other At the age of 46 years, pain in many joints, with swelling of the ankles.

At the present time, iritis of the right eye, severe pain in the left shoulder moderate dorsolumbar involvement and marked cervical stiffness. The rate of sedimentation is 20 mm (first hour) Roentgenograms of the sacroiliac joints *show bilateral and typical changes*

ONSET WITH INVOLVEMENT OF SEVERAL GROUPS OF JOINTS

The symptoms which we have described (Analysis of Symptoms, p 128) and grouped according to their spinal or peripheral localizations may be associated with one another and may form more or less complex pictures which escape all attempts at classification This mode of onset is less common than involvement of a single group of joints We have observed it in one fourth of the cases (69 out of 200)

FIRST EXAMPLE OF ONSET WITH SPINAL AND PERIPHERAL SYMPTOMS Mrs Er , aged 28 years. July 5, 1949 At the age of 23 years, six months after a trauma, attack of polyarticular inflammation with swelling of the small joints of both hands, and

at the same time general pain in the spine and pain radiating to the posterior part of the right thigh This attack was severe for one year, after which the patient continued to have sternal and sterno-costal pain, and slight exacerbations of pain in the ankles.

Now the patient presents a typical picture of Ankylosing Spondylitis

SECOND EXAMPLE OF ONSET WITH PAIN IN THE POSTERIOR PART OF THE THIGH AND IRITIS Mr L. O aged 53 years July 27 1929 At the age of 41 years, attack of iritis in the right eye, and at the same time pain in the posterior part of the left thigh progressive stiffening of the lumbar spine alternating with attacks of iritis each year for the past twelve years.

Now the patient presents a typical Ankylosing Spondylitis.

These examples illustrate the different ways in which, in a given patient, and from the onset of the disease, extremely varied symptoms may be grouped

Modes of Evolution

The symptoms which have just been analyzed vary greatly in their numerous localizations and in their features which, in a given peripheral joint or spinal segment, may be very different In order to outline the development of the disease, we have taken as a basic element stiffness of the spine, which has permitted us to divide the evolution of the disease into three periods phase of onset in which the stiffness is not marked and is confined to one segment the phase of obvious diagnosis or of definite stiffness, and the advanced phase in which the ankylosis is generally diffuse during this last phase, the inflammatory type of pain is usually absent, which indicates abatement in the activity of the process Now it seems advisable to describe the evolutionary features of spinal involvement, which are the classical elements of the disease, and their relation to peripheral involvement.

The disease progresses for many years, and it is a chronic condition in order to determine the treatment and the prognosis, therefore, it is essential to be familiar with the modes of evolution and to note and record the functional and articular symptoms after a period which we have arbitrarily set at twenty years

EVOLUTION OF SPINAL SYMPTOMS

Involvement of the spine is *often an early manifestation* (Modes of Onset, p 169) An analytical study of our cases has enabled us to specify three types of evolution in this situation

Ascending extension	90%
Simultaneous involvement of several segments or diffuse involvement from the start	7%
Descending extension (clinical)	3%

Ascending Extension

This was described by Strumpell (208) in 1884 as one of the features of the disease This mode of extension, in fact, is by far the most common we have observed it in 90% of our cases. Any spinal syndrome which, although at first it may be confined to the inferior segments, extends upward in the course of years, should be suspected of being due to Ankylosing Spondylitis

Table of the Date of Appearance of the Spinal Signs

(Statistics from 200 cases)

Years of Evolution	*Lumbar Involvement*	*Dorsal Involvement*	*Cervical Involvement*
First symptom	34 0	9 0	7
Up to one year	51 5	16 0	13
Up to 5 years	68 0	39 0	24
Up to 10 years	82 5	57 0	36
Up to 15 years	88 0	68 5	42
Up to 20 years	95 5	73 0	52
Persistent involvement ( )	87 0	64 5	66

( ) The reduction in the figures relating to the lumbar and dorsal segments is explained by the presence of resolving involvement.

In order to interpret the figures of this table, it is necessary to recall that the spinal signs are not always the first to occur

The spinal region most frequently affected at the beginning of the disease is the lumbar region, after the disease has progressed for one year, this segment is already found involved in more than 50% of the cases, while at the outset the dorsal and cervical segments are affected in about 10% of the cases [1]

The rapidity with which the upper segments become affected varies greatly in different cases

After the disease has progressed for one year, the frequency of involvement of the dorsal region is 16%, and that of the cervical region is 13%

After 10 years, a distinct increase in dorsal involvement up to 57% is observed During this phase of evolution, cervical involvement is more frequent, but its increase is less distinct 36%

[1] This figure of 10% corresponds either to cases with a diffuse spinal evolution from the start, or to cases with a cervical onset.

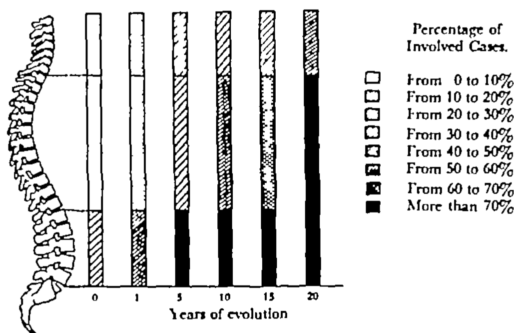


Figure 106 Diagram designed to illustrate ascending extension in Ankylosing Spondylitis. The data which permitted the preparation of this diagram were based on our statistics from 200 cases.

After 20 years, recent lumbar and dorsal involvement is rare, while cervical involvement continues to increase until its frequency reaches 66%

Diffuse and Early Involvement of the Spine

This was found in 7% of our cases, in three cases all three spinal segments were simultaneously affected

Forms of Descending Clinical Extension

In 3% of our cases in 2% the disease had begun in the cervical spine, and in 1% it had begun in the dorsal spine

RELATION BETWEEN THE EVOLUTION OF SPINAL AND PERIPHERAL INVOLVEMENT

We have already called attention to the fact that peripheral involvement may supervene at any phase of the disease, and that its relationship to spinal involvement varies greatly

1 *In 21% of our cases the disease had begun with peripheral involvement which had antedated the clinical signs of spinal involvement* (by from

6 months to 20 years (average, 7 years), that is, the disease had progressed for many years with purely peripheral clinical symptoms. Usually this peripheral involvement *progresses intermittently*, with long remissions.

In these cases the diagnosis is particularly difficult; at the outset some have been regarded as cases of rheumatoid arthritis. An accurate diagnosis was not made until the clinical signs of spinal involvement had developed.

2. In 11% of the cases the peripheral symptoms had appeared at the same time as the spinal symptoms.

3. In 40.5% of the cases peripheral involvement had developed during the progress of spinal disease.

TYPES OF EVOLUTION

In order to classify our cases we had to keep in mind not only the findings at the time of the first examination, but also those collected during the long years of observation. Thus, for the purpose of presentation, the different forms of evolution have been arranged in groups.

Table of Types of Evolution

Intermittent	47%
Intermittent, then continuous	16%
Progressive from the start	34%
Latent	3%

Intermittent Evolution

In nearly half of our cases (47%) the disease had progressed intermittently up to the time of the first examination. The disease had developed by peripheral or spinal episodes which had often occurred simultaneously. The severity of these attacks had varied greatly and their duration had ranged between two months and one year, or even two years.

The periods of remission, with return to normal of the sedimentation rate, may extend from a few months to several years. Attention should be drawn to the fact that remissions of five, 10, 15, and even 20 years, are relatively common.

During the remissions, physical examination may show a normal

condition or it may reveal more or less pronounced sequelae, and particularly a certain stiffness of the spine These sequelae often increase after each episode, but the patient, who no longer suffers from pain, regards himself as cured so long as he does not have marked functional impairment related to an extensive stiffness, or still more to a defective posture of the spine or of peripheral joints *But from what we have learned by observing our patients in whom* the disease had developed in widely spaced bouts, a patient who happens to be in a remission should be made to understand that the remission is only a temporary respite, and not a true cure

The intermittent recurrences of pain may continue for many years and throughout the course of the disease the ill-defined and benign forms often progress in this manner

Intermittent, and Later Continuous, Evolution

After the disease has progressed intermittently from two to 20 years, its progress may become continuous. In 16% of our cases the disease had progressed in this manner, and usually this had followed a severe episode in which several spinal segments had become affected

In a few cases this form of evolution seems to be related to deformities of the spine or peripheral joints

Continuous Evolution from the Start

Among our cases 34% have presented signs of continuous activity (pain, increase in deformity and increased sedimentation rate) In this category are found the severe forms (see Severe Forms) in which, in spite of a general ankylosis, the disease has continued to progress

STUDY OF CASES PROGRESSING FOR MORE THAN 20 YEARS IN WHICH THE PATIENTS HAD NOT RECEIVED REGULAR TREATMENT

This study is interesting from a prognostic point of view, because it can show us *the different modalities in the spontaneous course of the disease*

It is based on 40 cases, of which 33 were in men and seven in women, these figures corresponding to the proportion of our global statistics In these patients the disease had begun at approximately the same age as in the other patients included in our statistics

The period of evolution varied between 20 and 50 years (average, 24 8 years)

In only 12 cases, that is, in one-third of them, was the rate of sedimentation normal (less than 10 mm)

Benign or Ill Defined Type

Of this type there were 16 cases, or 40%

Spinal Symptoms

Stiffness of the spine was more or less extensive

Stiffness of all three segments	2 cases
Stiffness Lumbar and Dorsal	3 cases
Stiffness Lumbar and Cervical	2 cases
Stiffness of only one segment	8 cases
Stiffness absent	1 case

Kyphosis was always slight.

Peripheral Symptoms

On examination, most of these patients did not show any joint involvement in a few cases the involvement was slight.

Sedimentation Rate

It was normal in half of the cases, and in the others its increase was usually slight.

These patients had preserved a normal or quasi normal *occupational activity* Thus, even extensive stiffness of the spine when it is not accompanied by a defective posture of the trunk and by peripheral involvement, does not entail any noticeable degree of disability

In the course of their disease however a few of these patients had suffered *severe attacks* which had caused nearly complete, but temporary, disability To have given a grave prognosis at the time of these attacks would have proved an error Before giving

a prognosis, therefore, it is necessary to follow for a long time patients who are afflicted with Ankylosing Spondylitis.

A study of these cases shows that *in 40% of the patients suffering from Ankylosing Spondylitis the disease had a benign or ill-defined spontaneous evolution* this must be kept in mind if one is to judge the long term results of any method of treatment Our personal impression is that the percentage of benign cases is higher among the patients whom we have been able to follow from 10 to 15 years and who have been treated regularly

Type of Medium Severity with Partial Disability

There were 14 cases of this type or 35%

Spinal Symptoms

Stiffness of the spine was marked and very often diffuse

Stiffness affecting all spinal segments	11 cases
Stiffness affecting two spinal segments	2 cases
Stiffness affecting a single segment	1 case

In only one case did the patient have a *normal posture* and nearly always the anterior inclination of the head (occiput to-wall distance) was greater than 10 cm (12 cases), see the following figures

Normal posture	1 case
Kyphosis with occiput to-wall distance less than 5 cm.	1 case
Kyphosis with occiput-to-wall distance from 5 to 15 cm.	10 cases
Kyphosis with occiput to-wall distance greater than 15 cm	2 cases

Peripheral Symptoms

In eight cases the patients did not have any, or, when they were present, they were slight in four cases the patients presented more or less marked involvement of the hip In the two other cases slight involvement of several peripheral joints was present Except for the four cases with hip involvement, therefore, peripheral involvement was either slight or absent

Rate of Sedimentation

In most of the cases it was increased (11 out of 14) and it was normal in only three cases.

Our information on the *social activity* of these patients is incomplete they were not confined to bed A few could carry on occupations that were not tiring, such as that of punching tickets or that of watchman most of these patients did odd jobs at home

Severe Type, with Disability

Of this type there were 10 cases, or 25%

The proportion of these cases is much higher than the 10% indicated by our total statistics which included cases that had been seen at different times during the course of the disease The reason for this increase lies in the fact that not only the severity of the cases increases with the duration of the disease, but that it is chiefly the severe cases in which the patients come for medical care. They encumber hospital services while a certain number of benign or ill defined cases often escape attention at an ordinary casual examination

Spinal Symptoms

Stiffness of the spine is not much greater than in cases of medium severity in two of our cases the patients had only lumbar stiffness.

Stiffness of all three spinal segments	8 cases
Stiffness in only one spinal segment	2 cases

Nearly half of the severe cases had a marked *forward inclination of the head (occiput to-wall distance)* while an abnormal inclination was present in only one-sixth of the moderately severe cases, and none in the benign cases marked kyphosis, therefore, has a certain relationship to the severity of the disease As we have already mentioned we have observed in the cases with marked forward inclination of the head a pronounced loss of extension of the hips

Normal posture	2 cases
Kyphotic posture with occiput-to-wall distance less than 5 cm.	4 cases
Kyphotic posture with occiput-to-wall distance between 5 and 15 cm.	2 cases
Kyphotic posture with occiput-to-wall distance more than 15 cm.	2 cases

Peripheral Symptoms

In all the cases marked involvement of peripheral joints was present

The hips were affected in nine out of 10 cases, and in seven of them

the involvement was bilateral The hips were either "blocked" or greatly stiffened *in a faulty posture usually with flexion deformity*

The knees were affected in six cases, and the feet and ankles in two cases

Sedimentation Rate This was normal in three cases and increased in seven cases

Functional Capacity The constant involvement of the lower extremities made walking *very difficult or impossible* in five cases, that is, in half of them, the patients were confined to bed, and the five others moved with difficulty, either with crutches or walking sticks They were incapable of doing any remunerative work and they often needed help to dress or to take nourishment

In brief the spinal involvement in these cases was not distinctly greater than in cases of moderate severity *The gravity of these cases, therefore, came not so much from the spine as from the involvement of peripheral joints* Not one of our patients in whom only the spine was affected was an invalid From a functional point of view this brings out even more clearly the significance of peripheral involvement

Clinical Forms

Ankylosing Spondylitis is a general, chronic and protean disease which presents innumerable clinical aspects. We will describe only the aspects that have a sufficient individuality because of the absence of certain classical signs (the latent forms in which pain in the spine is absent) due to their benign or severe evolution, or because of the ' terrain in which the disease arises and progresses in women, old persons, and persons afflicted with psoriasis.

ANKYLOSING FORMS WITH PAINLESS SPINAL EVOLUTION

We designate as latent forms those which, *the disease having* progressed for more than 10 years, present a diffuse ankylosis of the spine, usually without important changes in statics, which has become established *without pain* and without the patient having become aware of it, at least as far as the dorsolumbar spine is concerned

From this category we have excluded cases with a latent evolution in one or two segments of the spine, in which the patients subsequently had pain in another segment, especially in the cervical segment.

Moreover we have noted many cases in which only a thorough interrogation of the patients disclosed the occurrence of short episodes of pain in the spine (Fig 27 bis) We have eliminated these from our study of the latent forms, but the clinical problem which they raise is the same as that of the true latent forms.

The peripheral involvement for which these patients had consulted a physician was often confined to the hips and knees rarely did they complain of intercostal pain These manifestations were similar to those noted in other cases of Ankylosing Spondylitis

In these patients only peripheral involvement raises therapeutic problems, because the spine usually is completely ankylosed in a satisfactory posture

In all the cases in which the sedimentation rate was determined it was normal or subnormal

Roentgenography of the spinal column shows extensive and pronounced lesions that produce a very peculiar picture (Figs 19 and 27) in three fourths of the cases, and in the remaining cases it shows less advanced lesions of the same kind

In antero-posterior roentgenograms the spine has a cylindrical and relatively homogeneous appearance, because the syndesmophytes are absent or only slightly prominent, the lateral contours of the spinal column, therefore, are not undulant as in the classical form in which the syndesmophytes jut out and produce the well known appearance of a bamboo stem, in these cases the vertebral bodies preserve their rectilinear borders The discs are often abnormally thin and appear to be ossified (Pathologic Anatomy, p 274)

Along the roentgenologic shadow of the column *three parallel rails* stand out a central rail caused by ossification of the inter spinous and supraspinous ligaments, and two other lateral rails due to ankylosis of the interapophyseal articulations and probably also to ossification of the ligamenta flava

The bone structure is such that the images have a ' washed out'' appearance

In all the cases the sacroiliac joints are completely fused (stage III), which is normal at this advanced phase

These cases are surprising because this diffuse ankylosis and the marked anatomic changes revealed by roentgenograms have appeared without having been preceded by painful manifestations in the spine.

We do not overlook the fact that cases occur in which a segmental involvement of the spine may remain painless and may not show the roentgenographic features which have just been described, but the fact that they are present in most of the latent cases leads us to think that this peculiar roentgenographic picture may be related to the absence of pain

ILL-DEFINED OR RESOLVING FORMS

Here we are dealing with patients who, after the disease has been active a long time (we have included only patients who have been afflicted more than 20 years), present only a slight limitation of spinal motion and in whom peripheral involvement has been slight or resolving The sedimentation rate is normal, the general condition is good, and the patients physical activity is normal (Figs. 107, 108a, b and c, also Fig 67)

These cases are relatively rare Their study which has also been carried out by Knutson presents a double interest because

A normal or practically normal mobility, when the spinal column is examined must not lead one to discard the diagnosis of Ankylosing Spondylitis, even after many years of evolution and because of the possibility of a spontaneous, benign or resolving evolution of Ankylosing Spondylitis

SEVERE ACTIVE FORMS

These forms are characterized by marked functional impairment which reduces the patients to a bedridden existence or nearly so (Fig 109), and by a continuously active disease which often seriously affects the general condition and which persists in spite of all therapeutic measures

This clinical picture has been observed 20 times out of 200 cases, or a proportion of 10% this proportion increases to 25% in the group of 40 cases in which the disease had been active more than 20 years (page 190)

Because of pain stiffness and deformity of most of the *peripheral joints* and especially those of the lower extremities, these patients are confined to bed all the time or half the time

Muscular atrophy is diffuse and pronounced and it also contributes to the patients disability

Because of the pathologic process itself and because of the prolonged confinement in bed the general condition of these patients is very precarious they are emaciated the skin is dry pale, and yellowish, and the nails undergo changes indicating a very bad trophic condition Digestive function is sluggish and deficient,

Figure 107 Figure 108a Figure 108b



Figure 108c

Figure 107 Same patient as in Figure 66 As may be recalled this patient presented an Ankylosing Spondylitis of 34 years duration. Photograph showing the normal statics of his spine.

Figure 108 Same patient as in Figure 107 the mobility of the spine may be regarded as normal (the patient's age was 51 years) a Lateral flexion. b Hyperextension c. Flexion.

and respiratory function reveals its insufficiency when the patient tries to make the slightest effort tachycardia and cyanosis of the legs

Sometimes the ocular complications that follow the recurring attacks of iritis cause a high percentage of the patients to become blind or half blind, and this increases the impression of physical decay

The rate of sedimentation always remains abnormally high, and sometimes greatly so 60 to 100 mm at the first hour in spite of medical treatment.

This clinical picture may develop either *rapidly from the start* or in other cases after *intermittent and repeated attacks* which have gradually become more and more severe

In these cases the therapeutic problem raises serious difficulties physical treatment, hot spring treatment, and treatment with *roentgen rays often are not well tolerated* internal treatment is not effective and often gives rise to reactions or incidents orthopedic correction *whether surgical or orthopedic*, is made difficult by the condition of the skin, by muscular atrophy very frequently by osteoporosis in bedridden patients, and by the involvement of many joints, and especially by the continuous activity of the disease without any period of remission that would be necessary for an operation

On the one hand an attempt should be made to improve the patient s general condition and on the other hand every effort should be made to prevent the aggravation of deformities, and the condition of the skin should be closely watched moreover radioactive agents (Thorium X) and now hormonal treatment (Cortisone, A C.T H) should be tried

EXTENSIVE PERIPHERAL FORMS

There is a severe form of inflammatory rheumatism which involves all the spinal or peripheral joints and ends in ankyloses By J Forestier this form has been individualized under the name of 'ensheathing panarthritis,' and vas studied in Vuillermet s thesis (Lyon, 1940) (214) e French edition of 1951



Figure 109 Mr G aged 46 years. Severe Ankylosing Spondylitis which has been active for 22 years complete ankylosis of the spine involvement of a large number of peripheral joints, with marked stiffness and deformity the patient is bedridden. Extreme muscular atrophy distinct impairment of visual acuity due to attacks of iritis.

as a severe form of Ankylosing Spondylitis Since then, two of us (237) have come to think that, in young persons, the polyarticular arthritis may involve not only the joints of the extremities, but also the sacroiliac joints and the interapophyseal joints, notably those of the cervical region they produce bony ankyloses of the same type as those seen in Ankylosing Spondylitis, but in them we have not found the sacroiliac changes of stages I and II, nor the bone bridges opposite the discs or overlying them, which appear to be characteristic of Ankylosing Spondylitis Therefore, this severe and diffuse polyarthritis should be distinguished from Ankylosing Spondylitis with extensive peripheral manifestations

In this form the *clinical picture of peripheral involvement* is extremely

severe, and it often begins in this manner severe pain, early anky losis of joints, and rapid muscular atrophy the general symptoms, fever and emaciation, are very pronounced

This picture of peripheral involvement tends to mask the progress of the clinical signs of spinal involvement, which are often veiled The patients are soon immobilized and confined to bed After they have suffered a series of successive attacks, these patients present complete bony ankylosis of most of their joints and marked atrophy of muscles The temporo-maxillary joints are often affected and eating becomes difficult, thus increasing the emacia tion besides this, we have noted fever as a sign of general disease During the period of obvious diagnosis, the patients can be truth fully described as "wooden men" because of their complete rigidity Because the patients so rapidly become bedridden, a fact which may also be responsible for the relatively slight pain in the spine vertebral ankylosis is not accompanied by marked kyphosis

During the terminal phase, trophic disturbances of the skin, hair and nails, edema and emaciation when eating is interfered with by ankylosis of the jaws, are likely to supervene. One of our patients died of asphyxia in the course of an ordinary laryngo-tracheitis because he could not expel the mucus.

The most characteristic *clinical element* is the appearance, in the knees and less often in the other peripheral joints, of an '*armor plating*' of the affected joints the peripatellar culs-de-sacs have lost their suppleness and have been replaced by a hard, resistant and ligneous layer devoid of elasticity which may be compared with old indurated rubber every part of the affected joints is enclosed in a resistant sheath (214) Palpation and attempts at mobiliza tion of these joints are extremely painful, even when they are anky losed

Roentgenograms of these ankylosed articulations show very fine linear, parallel strands thrown across, like bridges, from one epiphysis to another, but they are often incomplete The combina tion of all these strands forms a sort of veil which covers the articula tion The articular space often preserves a good width in spite of the linear strands, the articular space is often readily visible but sometimes it becomes opaque because of ossification of the articular cartilages (in one case this was verified surgically)



Figure 110 Same patient as in Figure 101 Extensive peripheral form roentgenogram of the pelvis picture of bony ankylosis of the hips in an inconceivably deformed attitude picture of ankylosis of the interapophyseal joints (noteworthy in the lumbar film, is the absence of syndesmophytes) ankylosis of the pubic symphysis diffuse osteoporosis.

In the extensive peripheral form these roentgenographic images have been observed especially in the hips (Fig 110), in the knees (Fig 93), *and much less frequently in other peripheral joints* (Figs 111, 112, 113 also Fig 101) These pictures present *the same features as those found only in the hips of typical cases of Ankylosing Spondylitis In the extensive peripheral form the diffusion of the ankylosing process is at its maximum*

PSORIATIC SPONDYLITIS

A distinctly individualized variety of Ankylosing Spondylitis develops in patients who have been afflicted with psoriasis [1] Of this

[1] A report on Psoriatic Rheumatism was published by Pinol, Aquade, Barcelo and Rotes-Querol (156).

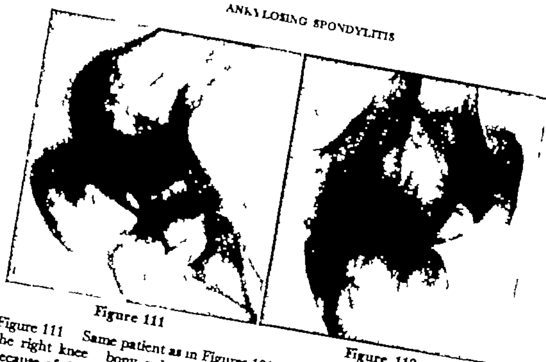


Figure 111 Figure 112

Figure 111 Same patient as in Figures 101 and 110 Lateral roentgenogram of the right knee bony ankylosis with normal thickness of the articular space because of the severe peripheral manifestations, the patient was bedridden from the start.

Figure 112. Mr Mic aged 34 years, with Ankylosing Spondylitis of 20 years' duration extensive peripheral form right knee involved from the beginning Lateral roentgenogram of this joint picture of bony ankylosis with disappearance of the joint space, and diffuse osteoporosis.

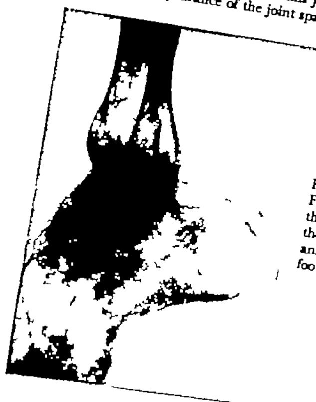


Figure 113 Same patient as in Fig 112. Lateral roentgenogram of the right ankle and foot affected by the disease from the beginning bony ankylosis of the different joints of the foot, with very marked equinism pronounced osteoporosis.

variety we have observed 13 cases *which were not included in our statistics bearing on 200 cases* The proportion of men and women has been about the same as in spondylitis without psoriasis 12 men and one woman

The age of onset varies between 12 and 51 years, the average age being about 30 years.

Clinical Considerations

Cutaneous Syndrome

The skin lesions do not have any clinical or anatomo-pathologic peculiarities that would enable one to distinguish them from those of ordinary psoriasis.

However, there are 1) a marked tendency to atypical lesions, but this tendency cannot well be described 2) a greater frequency of psoriatic involvement of the scalp and nails, and 3) a greater resistance to treatment of the cutaneous lesions

Chronologically the psoriasis usually antedates the articular involvement, as in rheumatoid arthritis this was noted in six of the eight cases in which it was possible to record the time of appearance of the two kinds of involvement in the two other cases the onset was simultaneous In the course of the disease the cutaneous and articular *exacerbations* have most frequently been concomitant, and more rarely alternating

Articular Syndrome

Modes of Onset

Six times the onset was purely peripheral

Three times the onset was purely spinal,

Four times peripheral and spinal symptoms were associated

Peripheral Articular Syndrome This is characterized by its frequency, its extension, and the sites of involvement. Peripheral involvement, which persisted at the time of the first examination was present in all the cases, while it was present in only 51% of the cases of Ankylosing Spondylitis without psoriasis

In our series of 13 cases peripheral involvement extended diffusely (Fig 114a) We have noted the nearly constant involvement of the shoulders, hips, and knees. The hands are involved in more

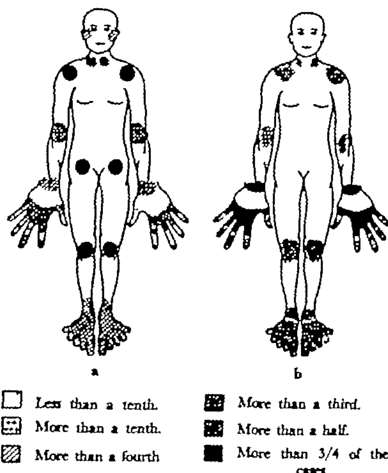


Figure 114 Distribution of peripheral articular involvement in a. Ankylosing Spondylitis with psoriasis. b Rheumatoid arthritis with psoriasis.

than half the cases (8 cases) *involvement of the distal phalangeal* joints, which was mentioned by Hench, is frequent (5 cases out of 13) and very characteristic its presence should lead one to look for psoriasis. Disturbances of the nails are often associated with this distal involvement. In one case there occurred an iritis identical to the iritis that occurs in ordinary Ankylosing Spondylitis.

From a functional point of view these forms of involvement are grave because of their generalization and their localization in important joints hips, knees, and shoulders, as well as because of the marked stiffness or ankylosis

The functional capacity of these patients is extremely reduced one third of our patients were completely bedridden one third

were semi invalids, that is, they could move with difficulty, and only one third preserved a certain degree of functional capacity which permitted them to be independent and to do a little work

Spinal Involvement Involvement of the spine with a clinical cervical onset and downward extension was noted in the history of five cases, with the reservation that the lumbar and dorsal involvement may have been latent or may have gone unperceived by the patient as well as by the physician because of the importance of the peripheral disturbances, examination of the spine of a bedridden patient is often difficult and the stiffness and the pain points over the spine may go unrecognized

It would be interesting to know if there is in these patients, any relation between the psoriatic involvement of the scalp and the cervical onset of the disease, which is more frequent than in ordinary Ankylosing Spondylitis

As in the case of involvement of peripheral joints, involvement of the spine rapidly becomes diffuse in 12 out of 13 cases all the spinal segments were affected at the time of the first examination

Roentgenology

Practically speaking the sacroiliac and spinal changes are in no way different from those observed in ordinary spondylitis, but because of their rapid progress, they are often pronounced fusion of the sacroiliac joints (stage III), numerous syndesmophytes, and lateral rails produced by ankylosis of the interapophyseal articulations (Fig 115)

The pelvis also may show important alterations in three cases the pubic symphysis was fused, and in three other cases marked proliferation of bone in the ischia was present

In one patient the structure of the entire pelvis had an altogether peculiar appearance with readily visible linear strands against an osteoporotic background, and these strands criss-crossed in all directions.

Besides the bony ankylosis with fine linear strands in the region of the hips (Fig 115), there may also be observed changes in the joints of the wrists, hands ankles and feet, changes which are not found in ordinary Ankylosing Spondylitis without psoriasis



Figure 115 Mr Vin aged 36 years, with Ankylosing Spondylitis and psoriasis cutaneous syndrome began seven years ago onset of the articular syndrome five years ago Roentgenogram of the sacroiliac joints and of the lumbar spine shows changes of the same kind as those occurring in ordinary Ankylosing Spondylitis. Noteworthy is the bony ankylosis of the hips with an articular space of normal width.



a



b

Figure 116 Mr Lor aged 47 years, with Ankylosing Spondylitis and psoriasis of 16 years duration simultaneous onset of the cutaneous and articular syndromes. a. Roentgenograms of the second, third, and fourth fingers of the right hand. b Roentgenogram of the left thumb Findings notches, phalanges pointed "like a pencil" or "in the form of a cupule, as well as their distal localizations, are characteristic of psoriatic rheumatism.

In the joints of the hands, where the changes have been carefully studied, they are peculiar because of their *early involvement of the distal phalangeal joints* while in ordinary rheumatoid arthritis involvement of these joints occurs much later Very characteristic *marginal notchings* as if produced by a finger nail indent *the border of the phalangeal epiphyses*, either in the zone contiguous to the joint or at a short distance from it. As for the epiphyses and the vicinity of the articular spaces, small, well circumscribed, rounded and *hollow masses or areas* are often visible, and these, like the erosions, may be an element of the osteolysis.

When the erosions of the epiphyses (Figs. 116a and b) become much more pronounced, they give to the epiphyses an irregular contour the epiphysis of the distal phalanx facing the proximal phalanx gradually becomes hollow and produces the 'cup image mentioned by American writers, while the other extremity becomes more pointed and produces the pencil point image. The two phalanges are then separated by a wider space than the normal articular space In contrast to the observations which have been made in rheumatoid arthritis, we have found in these pictures, not an image of osteoporosis, but on the contrary a slight increase in density of the bone structure bordering on erosion Subluxations and luxations are frequent

A more severe degree of bone change is produced by a marked osteolytic process which extends beyond the epiphysis and invades the diaphysis, of the phalanges only fragments may remain These are mutilating forms the joints no longer exist and the articular segments dangle and their length has diminished (opera glass fingers)

The *carpus* and *tarsus* show destruction of their articular borders, with fusion of all the bones and a greatly modified structure often including many small, circumscribed, rounded and hollow areas.

Summary

In patients who are afflicted with Ankylosing Spondylitis and psoriasis the spinal symptoms are similar to those of ordinary Ankylosing Spondylitis On the contrary the acromelic joints, especially the distal phalangeal joints which, in ordinary Ankylosing Spondylitis, are exceptionally involved are frequently affected

in these cases In these joints peculiar roentgenographic signs are observed marginal notchings, epiphyseal erosions, and mutilations In our limited series of 13 cases peripheral involvements were numerous and diffuse and most of these cases had to be classed among the severe progressive forms, sometimes with greatly increased sedimentation rates

Difference between the Psoriatic Form of Ankylosing Spondylitis and the Psoriatic Form of Rheumatoid Arthritis We have been struck by the distinct clinical difference between the cases of inflammatory rheumatism with spinal involvement and those in which the spine has remained clinically unaffected

In the psoriatic forms of chronic rheumatism, therefore, we can keep to the usual separation of the two main groups of rheumatic inflammations Ankylosing Spondylitis and Rheumatoid Arthritis

It is true that, in patients who are afflicted with psoriasis, some of the features of these two groups are modified

1) Modifications common to both groups
   Early involvement of the distal phalangeal joints
   Roentgenographic changes in the hands and feet marginal notchings, erosions,
2) Modifications peculiar to Ankylosing Spondylitis
   Greater extension and degree of peripheral involvement than in Ankylosing Spondylitis without psoriasis,

The fact that an inflammatory rheumatism, either Ankylosing Spondylitis or Rheumatoid Arthritis, should supervene in a patient who is already afflicted with psoriasis gives to these diseases additional features, but it does not erase the essential clinical and roentgenologic features by which they can be recognized The serologic tests that give different results in Ankylosing Spondylitis and in ordinary multiple arthritis may perhaps furnish arguments in favor of the co-existence of two different diseases in patients with psoriasis.

Therapeutic Deductions Classically, treatment with Gold salts should be closely supervised because of the possibility of erythrodermia, but we have never observed it Treatment with strong doses of Copper salts has given us some good results, and sometimes unexpected successes Thorium X also gives inconstant results. Treatment with hormones or with Cortisone and A C T H

has a rapid, but transitory, action on the cutaneous lesions as well as on the articular element. For the skin lesions as well as for the rheumatic disturbances hot springs treatment preserves its traditional indications

ANKYLOSING SPONDYLITIS IN OLD PERSONS

In aged persons two groups of Ankylosing Spondylitis can be distinguished

Those which, *after the disease has progressed* more or *less* latently until a certain age, subsequently present an attack of pain in the spine or in peripheral joints Cases such as these were recently described by M P Weil and Sichère (217) under the name of rheumatic spondylitis with a slow evolution and a late manifestation

These forms can be recognized by the disproportion between the short duration of clinical progress and the severity of the roentgenographic symptoms, moreover, sharp questioning often discloses peripheral or spinal symptoms of short duration which had developed long before the recent symptoms that attracted the attention of the patient.

The second group is composed of cases in which the disease really began at an advanced age (more than 50 years) these are very rare (3%) and they do not present any clinical peculiarity that would permit them to be distinguished from Ankylosing Spondylitis in young men Here we find the same spinal and peripheral *evolution, but the roentgenographic pictures show slight differences* from those seen in the usual form the sacroiliac lesions are always slight.

In stage I the zones of condensation practically do not exist, and the fusion of bones (stage III) is not complete In three cases there was an increased density of the upper third of the articular space corresponding to the fibrous part of the joint, but the lower two-thirds were still easily visible

Given the frequency of disc degeneration after the age of 50 years, the syndesmophyte curves around the protruding disc (Figs 28a, b, and c) and assumes an atypical appearance which is difficult to interpret One must look carefully for syndesmophytes

that present the usual features which may be found in other vertebral spaces in which the discs have undergone less degeneration

Another diagnosis that may be made is senile ankylosing vertebral hyperostosis of Forestier and Rotes Querol (p 231)

ANKYLOSING SPONDYLITIS IN WOMEN

In our general statistics from 200 cases 18% of the patients were women, but our study of female cases is based on 50 case records and therefore, includes 14 additional cases.

In women the *age of onset* of the disease is the same as in men (Curve of the Age of Onset, p 10)

An *analysis of the symptoms* does not reveal any significant differences between women and men

Spinal Symptoms

Statics of the Spine Spinal deformities are less common in women than in men and they are also less pronounced

Among 50 women lumbar lordosis had disappeared in 18 cases, dorsal kyphosis was exaggerated in 22 cases, and a flat back was observed in 13 cases All these female patients, except four, had an occiput to-wall distance less than 10 cm It may be recalled that in men, on the contrary, marked or very marked increases in the occiput to-wall distance are common

In women the lower frequency of disturbances in spinal posture and their moderate severity may be related to female morphology and the rarity among them of occupations requiring heavy labor

Downward extension of the disease also is as exceptional in women as in men

Peripheral Symptoms

Thirty nine out of 50 of our female patients had peripheral involvement, while 56 out of 100 of our male patients were similarly affected Statistically this difference is not significant. In both sexes the type of peripheral involvement is the same

Roentgenographic Features

In this respect we have not observed any distinct differences between men and women

Several of our patients have had a normal pregnancy and con finement (p 251)

The diagnosis is often more difficult in women than in men When faced with inflammatory rheumatic manifestations in women, the diagnosis of rheumatoid arthritis should receive special con sideration In women, moreover, the spinal deformities are less marked, and a systematic examination is often necessary to bring them out.

Nevertheless and after this study we are led to conclude that, in spite of the sexual differences as these may be affected by the age of puberty, the anatomy of the pelvis, and the type of respiration, we have not observed, between the cases of Ankylosing Spondyl itis occurring in men and those occurring in women, any differences sufficient to justify the description of a feminine form of the disease.

Diagnostic Problems

RULES TO BE FOLLOWED TO AVOID ERRORS

The clinical picture of Ankylosing Spondylitis in the phase of onset includes, in two-thirds of the cases, symptoms localized in the spine or in peripheral joints, or pain corresponding to the distribution of the nerve roots (Modes of Onset, p 169)

These symptoms are extremely varied and, by themselves, they present few features, or none, that would permit one to relate them to Ankylosing Spondylitis for example, the symptoms found on examination when the knee is involved may be the same whether the patient is afflicted with a rheumatic disease or with an early tuberculous infection Generally the diagnosis is oriented by the frequency of this or that disease in a given joint when the knee of a young individual is inflamed, one immediately thinks of a tuberculous infection but, as we have tried to show in this book, inflammatory rheumatism, and especially Ankylosing Spondylitis, is one of the frequent causes of inflammation of the knee therefore, this possibility also should be considered systematically and, *in every patient who presents any kind of articular involvement, the following steps should be taken*

1) Carry out a *complete examination of all the other joints and especially of the spine*

The patient should be examined without any clothing Each of the peripheral joints should be inspected for swelling, deformity, and vicious postures, palpated (sensation of fluid, synovial proliferation, and search for painful points), tested for mobility first by active motion (the patient himself shows what he can do), and then by passive movement (range of motion and pain induced by this motion)

The spine is first examined with the patient in a standing position The posture of the spine should first be carefully noted from the side, from the back, and from the front then the mobility of

each segment should be ascertained and recorded Then, by palpation with the patient in the prone position, the presence of painful points in the spine and the sacroiliac joints should be ascertained, these joints should also be examined by maneuvers designed to determine their mobility The clinical examination should also include testing the reflexes of the lower and upper extremities

Determination of the sedimentation rate should be made in every case In these patients it is important to carry out systematically the sheep cell agglutination test

Perhaps it may seem useless to repeat so often the importance of systematic examination, but our experience has unfortunately proved that, when such an examination has been neglected by us or by others, shocking errors in diagnosis have been committed, because Ankylosing Spondylitis is a localized disturbance only at the very beginning of its evolution In fact, *the disease rapidly becomes a diffuse process* when fresh symptoms are pronounced, the patient himself will call attention to them, but when they are slight, only a complete physical examination can reveal them

2) Moreover, Ankylosing Spondylitis has the peculiarity of revealing its presence by *an extremely early involvement of the sacroiliac joints* which, until now, should be considered present from the very onset of the disease and which expresses itself roentgenographically by characteristic pictures, according to one of us, this is the 'key' to early diagnosis Also *in every young patient who presents an articular syndrome, when physical examination does not permit an accurate etiologic diagnosis, the local and general examination should systematically be completed by standard roentgenograms of the sacroiliac joints* Later the association of different symptoms (hydrarthrosis and iritis, lumbago and tendinitis)—an association which is characteristic of Ankylosing Spondylitis—will lead from the outset to the diagnosis of Ankylosing Spondylitis, and roentgenography of the sacroiliac joints *will only serve to confirm the clinical diagnosis*

During the discussion of the diagnoses that may be made during the *phase of onset*, we will continue to repeat over and over again the two following points systematic clinical examination and roentgenography of the sacroiliac joints With these elements the diagnosis will usually appear evident.

Sometimes, however, the problem may be more complex, either because of the *co-existence of two common diseases* (Scheuermann's disease and Ankylosing Spondylitis), or because of the finding of spinal anomalies (sacralization lumbalization, etc) Therefore, the temptation to make too rapidly a "roentgenographic' diagnosis based on obvious pictures of a common disease or of some anomaly should be avoided, a systematic interpretation of the films often enables one to discover less obvious changes which may be characteristic of Ankylosing Spondylitis, but which may be 'masked' by images that had caught the interpreter's attention during a rapid reading

When the patient is seen during the phase of onset, which, it must be remembered may be quite long, two elements immediately help in orienting the diagnosis these are, on the one hand, *the very peculiar mode of evolution of Ankylosing Spondylitis,* of which the chief feature is *the intermittence of its manifestations,* whether these be due to peripheral or spinal involvement (see Modes of Evolution), and on the other hand, the *multiplicity of the affected joints* Little by little, in fact, new sites of involvement have appeared, therefore, the process is not a local disease, but a general one. One point which must be stressed is that inflammatory rheumatism is the most common diffuse articular disturbance, while tuberculosis, which is one of the great classical causes of articular disease, rarely develops *in many different sites* and thus it forms a sharp contrast to Ankylosing Spondylitis

During the phase of obvious diagnosis, and with still greater reason during the advanced phase, as these have been defined the patient presents a distinct and extensive stiffness of the spine, this stiffness either has been mentioned by the patient because of the pain which accompanied its development, or has been found in the course of *a systematic examination* of the spine when the stiffness had developed without pain

In *adolescents or young adults* this picture of spinal stiffness, with the usual defects in posture that accompany it, does not raise any diagnostic problems, because not only the clinical signs are typical but, in roentgenograms, the syndesmophytes and sometimes the interapophyseal articular changes are present In *aged persons,* on the contrary it is sometimes difficult to make a positive diagnosis

of Ankylosing Spondylitis because, owing to the antecedent condition of the discs, the syndesmophytes are found superimposed on the osteophytes, and because of this, they do not have their usual features, it is therefore necessary to have made

1) Roentgenograms of another spinal region that may reveal typical syndesmophytes

2) A roentgenogram of the sacroiliac joints which, save in altogether exceptional cases, show typical lesions.

In old persons, moreover, there exists an ankylosing disease, the senile ankylosing hyperostosis of the spine of J Forestier and Rotes-Querol which, thanks to its roentgenographic signs, may be distinguished from Ankylosing Spondylitis.

DIAGNOSTIC PROBLEMS IN THE PHASE OF ONSET

We have shown that Ankylosing Spondylitis at the beginning is a disease that may manifest itself by a single group of symptoms. We will now study separately the different groups of initial symptoms, and in connection with each group we will describe the diagnostic elements supported by a reading of the roentgenographic films

Spinal Symptoms

Lumbar Segment

Because of the absence of roentgenographic changes in the lumbar spine that are characteristic of the disease the clinical manifestations during the phase of onset are interpreted as symptoms of an infectious disease (Pott's disease) or of a static disturbance

Pott's Disease (Tuberculous Spondylitis) In adults and even in children this disease may present a latent roentgenographic phase (1 or 2 years), and then slight roentgenographic signs may sometimes develop over a rather long period So it is necessary to specify the character of its first clinical and roentgenographic symptoms

Differential Clinical Signs *Pott's disease sometimes begins* by a period of invasion with general signs, fever and diffuse pain Exceptionally, a similar picture may be observed in Ankylosing

Spondylitis, but in the first case the symptoms rapidly become localized in one spinal focus

Clinically the differential diagnosis that should be made between Pott s disease and Ankylosing Spondylitis in their early stages is that of a spinal "focal" disease and of a diffuse vertebral malady

The following symptoms favor the diagnosis of Pott's disease

Localization of pain on pressure over one or two vertebrae It is known that, in Ankylosing Spondylitis, pain induced in a single vertebra is rare usually it is diffuse or localized at many points (Table of Pain Points, Fig 4)

The character of the pain caused by pressure over the spinous processes is often very dull and is quite different from the acute pain of Ankylosing Spondylitis

The marked segmental stiffness with muscular contracture differs greatly from the diffuse stiffness of Ankylosing Spondylitis

Since the rate of sedimentation may be increased in both diseases, it is not useful for the differential diagnosis

Usually *the first roentgenographic symptom* of Pott s disease is thinning of a single intervertebral disc in Ankylosing Spondylitis thinning of a disc may be observed, but it usually appears late and then it is found at several levels.

In the phase of onset the other infectious forms of spondylitis (staphylococcic or streptococcic) do not raise any diagnostic problem the clinical picture of these conditions is very striking and the patient is obliged to go to bed It should be recalled that these infectious forms of spondylitis, which progress rapidly, yield pictures of syndesmophytes affecting one or two pairs of contiguous vertebrae (Fig 17 bis), which should be distinguished from the syndesmophytes in Ankylosing Spondylitis.

Static Pain in Young or Adult Patients At the beginning the spinal symptoms presented by a young patient who is afflicted with Ankylosing Spondylitis may be atypical

The pain is completely relieved by sufficiently prolonged rest, but it may reappear when the patient stands or when he exerts any effort.

Definite stiffness is absent in the standing position, and para vertebral contracture is absent when the patient lies down *the diagnosis of static origin* of the pain is envisaged and seems *to be con*

firmed by roentgenography signs of loss of static balance (asymmetrical pelvis, increased lordosis, scoliosis, spondylolisthesis of the isthmus[1]), and transitory anomalies (sacralization, lumbalization, spina bifida)

But when the usual treatment of these static difficulties does not yield satisfactory results, the patient should be completely re examined that is, the examiner should analyze anew the character of the pain which may have changed and may have become more characteristic of Ankylosing Spondylitis, and the patient should be re-examined Roederer's *phrase may be cited in this connection* "Here is a sacralization, now let us seek the cause of pain '

Dorsal and Cervical Segments

Practically speaking, as we have already seen, *these localizations had not presented themselves as isolated phenomena* When pain in the dorsal region has made its appearance we have always found stiffness in the lumbar portion of the spine, of which the patient may not have been aware. Too rapid an examination of the spine may have allowed this stiffness of the lumbar segment to escape the attention of the examiner and may have led him to request only a dorsal roentgenogram If this had revealed certain changes, these might have been regarded as explaining the clinical symptoms and might have led to an error in diagnosis

These images are seen in Scheuermann s disease (187) which is usually localized between D-6 and D 11 and which is characterized by a wedge-shaped deformity of the vertebrae and an irregularity of the vertebral plateaux Intraspongiosal hernias of Schmorl, with reduction in the thickness of the discs, may sometimes be associated

For example, the diagnosis of Scheuermann's disease would seem quite natural especially when the young patient presents a kyphosis without the disappearance of lordosis, which sometimes occurs in Ankylosing Spondylitis

We have observed many cases in which Scheuermann s disease was associated with Ankylosing Spondylitis, or a calcified *nucleus pulposus* with Ankylosing Spondylitis. Roentgenograms should be

[1] Spondylolisthesis due to lengthening of the true isthmus this is a congenital anomaly of the lumbo-sacral transition zone.

tematically read, and one should bear in mind not only the most parent roentgenographic signs Although the rule should be to ribute different symptoms to one disease and not to several con ions, it should be remembered that some are so common that rtain associations may have occurred moreover one should ow that the pictures of ossified nuclei and of irregularities in the rtebral plateaux of the dorsal spine are very common and often not have any clinical significance *In the presence of such images,* erefore, *one should systematically seek some other cause for the pain com ined of by the patient*

These pictures may also represent a Kümmel Verneuil syn ome [1] breakdown of the plateaux, body wedge shaped or with creased density or an unrecognized image of fracture in one or veral spinous or transverse processes, on the anterior border the vertebrae The problem is the same as in the images of heuermann's disease with, in some cases, the additional factor traumatism on which the patient readily insists

Sometimes a differential diagnosis of wedge shaped vertebrae ust be made, because these images may be produced, not only by aumatism, but also by Ankylosing Spondylitis (Figs 32 and 33) the Kummel Verneuil syndrome there is often an irregularity one or more vertebral plateaux, results of the fracture, while in nkylosing Spondylitis the vertebral body presents only a wedge aped deformity due to its pathologic plasticity

Sacroiliac Symptoms

In this phase of onset only the roentgenographic changes in the croiliac joints permit one to make the diagnosis of Ankylosing pondylitis When the symptoms were analyzed, we described e features of these roentgenographic images, but they should be stinguished from those of other diseases when they are *slight* ey may be mistaken for early pictures of other conditions (sacro-oxalgia osteochondritis) when they are *marked*, but slightly typical, they may resemble the images of diseases such as sacro-oxalgia (pseudo-tuberculous changes of Ankylosing Spondylitis) or steitis condensans ilii.

[1] Post-traumatic spondylitis. This condition should be called "Kümmel-Verneuil-. Forestier syndrome."

When, in spite of clinical symptoms suggesting Ankylosing Spondylitis, the roentgenograms of the sacroiliac joints are normal, the patient should be regularly re-examined every three or six months (p 86)

Sacro-Coxalgia[1]

DIFFERENTIAL SACROILIAC SIGNS IN ANKYLOSING SPONDYLITIS AND IN SACRO-COXALGIA

Symptoms	*Ankylosing Spondylitis*	*Sacroiliac Tuberculosis*
Sex.	Most frequent in men	Equal frequency in men and women.
Clinical		
Severity of the sacro-iliac syndrome	Slight or moderate	Severe.
Abscess	Absent	Frequent.
Symptoms in other regions	Spinal or peripheral very frequent	Usually isolated sacro-iliac syndrome without associated symptoms.
Sedimentation Rate	Increased in 75% of the cases	Often increased.
Roentgenography		
Unilateral involvement	Rare and only at the beginning	Almost the rule.
Bilateral involvement	Constant, except at the outset;	Exceptional.
Bone destruction	absent (at most pseudo-widening and false, rounded areas with light centers)	Often very pronounced, marked loss of substance with eroded contours.
Juxta-articular condensation of bone	Very frequent	Very rare.
Separation of the pubic symphysis	Not observed	Frequent.

(Modified and completed from Broeher and Foerstler)

The roentgenographic images seen during the onset of sacro-coxalgia and of Ankylosing Spondylitis may be identical We have previously stressed the usually bilateral distribution of the sacroiliac changes in Ankylosing Spondylitis this bilateral distribution is very important, because we greatly doubt the existence of bilateral involvement in sacro-coxalgia S T Soholt (263),

[1] Sacroiliac tuberculosis.

however, claims to have seen bilateral involvement seven times in 75 cases of sacro-coxalgia

In rare cases of Ankylosing Spondylitis the lesions may be unilateral at the outset but when certain images are unilateral, they cannot be distinguished from those of tuberculosis pseudo-widening of the joint space, and irregular contour without definite subchondral condensation Then a differential diagnosis cannot be made, the clinical element has only a relative value, indeed, the symptoms of sacro-coxalgia, which are usually severe, may be moderate, and the sacroiliac symptoms of Ankylosing Spondylitis may sometimes be very striking In sacro-coxalgia it is the generally continuous *clinical progression* of the disease which, in Ankylosing Spondylitis, is intermittent, that is, interrupted by remissions, or it is the *roentgenographic progression* of the pathologic changes (unilateral images of destruction in sacro-coxalgia, and bilateral distribution and diffuse changes in Ankylosing Spondylitis) which make the diagnosis possible In such cases, therefore, it is wise not immediately to make the diagnosis of sacro-coxalgia and not to undertake a grafting operation, which was done in many cases which we subsequently examined

Melitococcic Sacroiliac Arthritis (254)

The usually striking clinical sacroiliac symptoms of this form of arthritis may not be marked in some cases Or the bilateral roentgenographic changes which, in Mediterranean fever are rather common are difficult to distinguish from those of Ankylosing Spondylitis In regions in which Brucellosis is endemic, therefore, it would be wise, in doubtful cases, to have serologic tests made to eliminate the possibility of Mediterranean fever

Osteochondritis or Sacroiliac Epiphysitis

In 1935 Marc H Rogers and Edwin Cleaves described in young persons, under the name of ' sacroiliac syndrome in the adolescent'' an osteochondritis localized in the sacroiliac joints Of this condition the clinical and roentgenologic features would seem to be those of a resolving sacroiliac syndrome without any characteristic symptom The 10 cases cited by these writers had been followed only for a few months The epiphyses affected by osteochondritis

would seem to be those that are situated in the outer part of the sacral bones and that are part of the sacroiliac joint (172)

These epiphyses, which develop late, are visible in roentgenograms from the fifteenth to the sixteenth year (Figs. 59a, b and c) and they become fused to the sacrum one or two years later (187) It is interesting to recall that the iliac bone does not have an epiphysis corresponding to the sacroiliac joint. In the description of cases of osteochondritis an anatomic verification to confirm the existence of involvement of the sacral epiphyses has not been made By all means this site of osteochondritis should be extremely rare, personally we have not observed a single case and, before making this diagnosis, we consider it wise to require that the following conditions should be fulfilled

a Age of the patient between 15 and 20 years, which is the period of ossification of these epiphyses

b Roentgenographic changes limited to the sacral portion to which the epiphyses belong

c Unilateral lesions, bilateral lesions in osteochondritis being an exceptional occurrence

d Observation of the patient for many years to make sure that the disease is not an Ankylosing Spondylitis in the period of onset.

Osteitis Condensans Ilii

Because this disease is not well known in France, it has seemed wise for us to describe its features the roentgenographic changes have been well set forth by Barsony and Polgar (7) and other writers have tried, by prolonged observation, to ascertain its evolution (Rendich and Schapiro in 1936 [166], Hare and Haggard in 1945 [100]) Although our personal experience is still very limited it seems to us that Ankylosing Spondylitis and *Osteitis Condensans Ilii* present distinct differential features

Always, it seems, the disease occurs in women (the male cases that have been cited by certain writers seem doubtful according to us and others, these are cases of Ankylosing Spondylitis in the phase of onset) The disease begins between the ages of 20 and 40 years most frequently the onset is related to pregnancy or confinement which, for certain writers (15) would even seem to

be constant factors, or less frequently to trauma, in a few cases an apparent cause cannot be found

Clinically it presents a sacroiliac syndrome which is often severe and consists of pain in the joint, and in half the cases the pain radiates to the posterior part of the thigh (100) Pressure over the sacroiliac joint spaces and attempts to mobilize these joints cause pain in the very region of the joints This syndrome is unilateral or bilateral, but it predominates on one side. Other writers have described latent cases (166) Examination of the spine yields negative results, and the patient s general condition is not greatly affected The rate of sedimentation seeems to remain normal

Roentgenography of the sacroiliac joints (Fig 117b) shows a uniform increase in density of the *iliac bone*, in the zone adjacent to the articular space This increase in density is bilateral and symmetrical, even in cases in which the clinical involvement is unilateral This zone of increased density has the form either of a rough isosceles triangle, with its base a few millimeters above the innominate line, at the upper extremity of the foot, or it has a rounded form The limit between the normal and abnormal parts of the bone is distinct. The superimposition of the innermost part of the iliac wing and of the sacrum makes it difficult to be certain whether the increased density belongs strictly to the ilium or whether it slightly involves the sacrum. In antero-posterior roentgenograms the lower part, or foot, of the articular space is always sharp The anterior or outer branch, on the contrary, which projects over the area of increased density is difficult to interpret. The posterior, or internal, branch, which often marks the inner limit of increased density, is distinctly visible. The articular space, which is visible in oblique views or in tomograms, seems never to be altered

Evolution The progress of the disease is either intermittent, with exacerbations lasting from several weeks to several months, or continuous with exacerbations. Its total duration may be very long, but we do not have any data from cases that have been followed long enough to have a definite idea In no case has an evolution toward ankylosis of the sacroiliac joints or toward extension to the spine been observed Shipp and Haggart have observed regression of the roentgenographic image



Figure 117 Sacroiliac changes in Ankylosing Spondylitis and Osteitis Condensans Ilii a. Bilateral sacroiliac changes in Ankylosing Spondylitis (stage I) greatly widened joint spaces with irregular contours and marked subchondral areas of condensation. Same patient as in Figure 33 b. Osteitis Condensans Ilii triangular zone of condensation situated in the middle portion of the iliac wing and adjacent to the sacroiliac joint space, the contours of which are well defined. Mrs. Gr aged 34 years, presents a left sacroiliac syndrome which has been present for three years and which followed a trauma this patient has been pregnant twice once six years ago, and the second time three years ago Examination disclosed a point of very sharp pain over the left sacroiliac joint every movement of the left lower extremity caused pain in the same joint.

The cases seen in obstetrical practice by James Young (221) during or after pregnancy, in which the symptoms consisted of sacroiliac or pubic pain, sometimes with an abnormal mobility of the ilia due to relaxation of ligaments, but without roentgenographic changes, should be followed in order to determine if the symptoms complained of by these patients might represent the onset of *Osteitis Condensans Ilii* which the rheumatologist will be called upon later to examine

From the point of view of pathologic anatomy, a report has been published with a biopsy of tissue removed during a grafting operation (166) This showed condensation of the bone trabeculae, independent of any neoplasmic or inflammatory process This condition seems to be refractory to ordinary methods of treatment, to roentgen treatment, etc

This disease affects the iliac bone, its evolution, therefore, differs from that of Ankylosing Spondylitis which, though for a time it is localized in the sacroiliac joints, may later become general

The roentgenographic changes occurring in osteitis condensans ilii have distinct features the articular space is not affected, the increased density of bone is homogeneous, and the contours of these areas of increased density are clean-cut (Fig 117b), and these appearances are quite different from those seen in Ankylosing Spondylitis When condensation bands border the sacroiliac joint spaces, Ankylosing Spondylitis should systematically be suspected

Sacroiliac Osteoarthritis

The question of differential diagnosis between images of sacroiliac osteoarthritis and the sacroiliac changes of spondylitis does not arise the sharp line of sclerosis due to senescence, limiting an articular space of normal width, should not be confused with the band of condensation bordering a greatly widened joint space in Ankylosing Spondylitis

Radiating Pain

Pain Radiating to the Lower Extremities

Monoradicular Sciatica. A study of deep referred pain in the buttocks and in the posterior part of the thighs, which has a sac-

roiliac origin, has made it possible for us to distinguish this kind of pain from pain of radicular origin

This root pain[1] (Symptoms of Onset, p 173) is rare, and as an "isolated symptom' it is exceptional, usually and from the outset it is associated with lumbar symptoms, the problem raised therefore is the origin of the lumbo-sciatic pain The most frequent cause of this pain is *protrusion of a nucleus pulposus,* or at least of an intervertebral disc But certain clinical features may lead one to make a diagnosis of Ankylosing Spondylitis

Pain chiefly at night

Absence of lateral flexion of the trunk,

Increase in the sedimentation rate,

Absence of roentgenographic signs favoring herniation of a nucleus pulposus (in sciatica from this cause these signs, in fact, are rarely absent [193])

But, as we have already mentioned in connection with the diagnostic problems in the lumbar region, *the tendency to ascribe* the lumbo-sciatic pain to over-exertion of an intervertebral disc should be avoided when an anomaly of the fifth lumbar vertebra has been disclosed

Deep Referred Pain In the buttock and posterior part of the lower extremity (practically speaking of the thigh)

This kind of pain is common at the outset (Analysis of Symptoms, p 85) Its features are pain in the buttock and posterior part of the thigh, sometimes extending lower, but never reaching the foot. It is a deep dull but usually moderate, pain which often appears during the night and which tends to increase especially when the patient sits for a long time, it is not accompanied by objective neurologic signs. This kind of pain should suggest a sacroiliac disturbance (diagnostic problems of the sacroiliac syndrome, p 91), and one should look for clinical symptoms and roentgenographic changes

Pain Radiating to the Thorax and Abdomen (*Pseudo-visceral Forms of H Forestier*)

For many years a few of our patients had consulted specialists

[1] Until now in Ankylosing Spondylitis we have observed only sciatic pain with an L-5 distribution. We have never seen a case with an S-1 distribution.

because of painful syndromes in the thorax and abdomen In some cases pleurisy, angina pectoris, or intercostal neuralgia had been considered, and in other cases the patients had been treated for a gastric disturbance, for cholecystitis, for appendicitis, or for uterine disorders.

Example of the pseudo-visceral form, with symptoms suggesting a renal disturbance

Mr D L. aged 40 years This patient had been suffering for five years His illness had begun with pain in the right hypochondrium, and this had been regarded as a sign of hydronephrosis, more recently the pain had radiated to the groin and right thigh. Then a complete roentgenographic exploration of the urinary system had given negative results. But two years later we found, on examination a marked lumbar rigidity and pain on pressure over the spinous processes of this segment We requested the privilege of inspecting the pyelographic roentgenograms that had been made two years previously and in these we recognized the characteristic sacroiliac changes of Ankylosing Spondylitis, although the spinal skeleton was normal.

Therefore, this was an example of those rather frequent cases in which the radiating pain is considered as forming part of a visceral syndrome.

Peripheral Symptoms

Involvement of the joints of the extremities is often the reason that leads the patient to consult a physician only detailed questioning, and especially a careful physical examination, can reveal stiffness or ankylosis of the spine when the disease is in the phase of obvious diagnosis, or slight signs of sacroiliac or lumbar involvement when the disease is still in the period of onset. But *at the very beginning of the disease* clinical involvement is strictly peripheral in a rather high percentage of cases (21%) The diagnostic problem varies with the number of affected joints.

Monarticular Arthritis

A tuberculous osteo-arthritis or an inflammatory involvement of rheumatic type may, at the outset, present the same symptoms pain swelling, muscular atrophy, and even absence of adenopathy,

and roentgenograms of the affected joint may show either a slight diffuse osteoporosis or a normal picture (because of roentgenographic latency) The fluid withdrawn by puncture may be identical from a chemical point of view (mucin, glucose) or from a cytologic point of view (40 175) Search for tubercle bacilli should be made, either by simple slides or by culture or inoculation of guinea pigs (this is positive in 84% of the cases) (121) In cases in which guinea pig inoculation yields a negative result, a surgical or orthopedic intervention that might result in ankylosis should not be undertaken, but the patient should be put to rest and the progress of his condition should be observed [1]

Sometimes, during this phase, not a single local element enables one to know the origin of the inflammatory arthritis and when systematic roentgenography of the sacroiliac joints is not done, only the evolution of the disease will make it possible to make a diagnosis From a therapeutic point of view, therefore, one must be extremely careful not to immobilize the joints in plaster as long as the clinical diagnosis has not been made It should be recalled that the first positive roentgenologic sign in favor of tuberculosis is the image of a destructive epiphyseal or juxta-epiphyseal focus.

When the diagnosis is still in doubt after the disease has progressed for several months, an articular biopsy is justifiable, but this biopsy should be made with certain precautions We lost a patient by generalization of tuberculosis after biopsy of the knee Since then, we systematically carry out immobilization in plaster for about ten days It is important to know that in cases of tuberculous arthritis, articular biopsy does not always give a positive result, because the tissue used for biopsy may have been removed from a non pathologic part of a diseased joint. The removed fragment of tissue should be implanted into the peritoneal cavity of the guinea pig which increases the number of positive cases When an adenopathy appears (this may supervene during an inflammatory rheumatism) a lymph node may be removed and examined histologically or still better a fragment of the node can

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[1] In one of our cases the synovial fluid which was inoculated into two guinea pigs did not produce any discoverable sign of tuberculosis in one guinea pig which was sacrificed after the classical interval of eight weeks, but the second pig presented symptoms at the end of five months and histologic examination revealed Koch's bacillus.

be implanted into the peritoneum of a guinea pig in an attempt to prove the presence of tuberculosis (Richard-Sotès)

Hydrarthrosis of the Knee

At the outset of Ankylosing Spondylitis, hydrarthrosis of one or both knees may, more than any other kind of involvement, remain "isolated" for many years, and its intermittent progression resembles that of other kinds of hydrarthrosis.

When it is unilateral, which is rather rare, the first attack, as a rule, suggests a traumatism and putting the patient at rest will be thought responsible for the disappearance of the symptoms But when it persists or recurs, and when it always remains unilateral, pain on movement and limitation of mobility suggest a diagnosis of meniscal lesions In the event that the results of exploratory puncture[1] and of pneumo-arthrography, when necessary, should not confirm such a meniscal origin, the diagnosis will then have to distinguish between the effusive form of tuberculous arthritis and effusion incidental to inflammatory rheumatism As long as the clinical or roentgenologic signs remain equivocal, the origin of the hydrarthrosis cannot be determined

But most frequently this effusive involvement becomes simultaneously or alternatingly *bilateral* When repeated attacks of bilateral disease occur and when the composition of the fluid obtained by puncture remains the same, an inflammatory origin must be considered but at the outset three rheumatic conditions may be localized in the knees and may manifest themselves by effusion chronic polyarticular hydrops (J Forestier), rheumatoid arthritis, and Ankylosing Spondylitis Only the evolution of the disease and the appearance of new sites of involvement can permit one to make the diagnosis [2]

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[1] In mechanical hydrarthrosis articular puncture (40 175) reveals a normal content of mucin and glucose and a slightly increased number of leucocytes (200 to 500 leucocytes) with predominance of lymphocytes, while in inflammatory diseases the mucin and glucose content is reduced and the number of leucocytes is slightly increased but with predominance of polymorphonuclears.

[2] At the present time we do not have records that would permit us to claim that, when only the clinical phenomena of hydrarthrosis in the knees are present, changes in the sacroiliac joints are or are not present. We do not know if the sacroiliac changes are present when only peripheral manifestations have developed.

Oligo-arthritides

We call 'oligo-arthritis' a generally asymmetric involvement of a few joints (5 or less) The oligo-arthritis of Ankylosing Spondylitis should be distinguished from that which may constitute the onset of a rheumatoid arthritis sex (female) age (more than 40 years) localization in the wrists and hands points toward rheumatoid arthritis The oligo-arthritis of Ankylosing Spondylitis presents two features 1) the *intermittence of its manifestations*, an intermittence which it is true, often disappears after a few attacks, when the disease progresses continuously and 2) its localization in the lower extremities.

Inflammatory involvement of the feet and the intermittent attacks are symptoms which can also be found in chronic polyarticular gout. When these symptoms develop in a *young man* (undeserved gout) one should systematically think of Ankylosing Spondylitis, but also of gout, and in favor of the latter one should look for family antecedents and slight clinical signs sudden onset of the attacks, tophi in the pavilion of the ear, and local signs A distinctly abnormal amount of uric acid in the blood will confirm the diagnosis

Attacks of Transient Polyarticular Inflammation

Every attack of polyarticular inflammation, especially when it occurs in a young person suggests the diagnosis of acute rheumatic fever Later on we will see the relations between Ankylosing Spondylitis and acute rheumatic fever

The transitory polyarticular exacerbations of rheumatoid arthritis and of Ankylosing Spondylitis may be differentiated from those of acute rheumatic fever by the early involvement of several joints, the greater frequency of localization in the phalangeal joints of the hands and feet and in the interapophyseal joints of the cervical spine the more uniform occurrence of these sites of involvement and the tendency to chronicity of one or several of them and the absence of cardiac signs But these attacks may be observed in the stage of onset of rheumatoid arthritis and in infectious pseudo-rheumatism. The fact that they occur in young men should attract attention because of the possibility of Ankylosing Spondylitis

Chronic Polyarticular Involvement Analogous to that of Rheumatoid Arthritis

From a practical standpoint this problem does not arise at the beginning it is only during the *advanced stage* of the disease that peripheral involvement suggesting the diagnosis of rheumatoid arthritis is observed, but systematic examination will reveal the

DIFFERENTIAL SIGNS OF ANKYLOSING SPONDYLITIS AND OF RHEUMATOID ARTHRITIS

(Based on 200 cases of Ankylosing Spondylitis and 100 cases of Rheumatoid Arthritis)

	Ankylosing Spondylitis	*Rheumatoid Arthritis*
Clinical Features.		
Age	Not observed at extreme ages	Observed at any age even in nursing infants and old men.
Sex	In 82% of men In 18% of women	In 20% of men In 80% of women.
Spinal involvement	Constant and extensive	Only in the cervical region in 20% of the cases
Peripheral involvement found at the first examination	In 72.5% of the cases	Constant.
Respiratory amplitude	Diminished in 91% of the cases	Always normal.
Distribution of peripheral involvement	Predominant in shoulders, hips, and lower extremities	Predominant in hands feet and skull.
Hands	Rarely affected and never systematically	Usually systematically involved.
Iritis	8.5% of the cases	In no case.
Subcutaneous nodules	Never observed	In 15% of the cases.
Roentgenologic Features		
Sacroiliac	Involved in all cases	Never involved.
Syndesmophytes	Very frequent	Never observed.
Interapophyseal arthritides	Frequent	Cervical region in only 20% of the cases.
Other features distinctly visible in the hips	Evolution toward bony ankylosis	Bone destruction (often with acetabular fusion) Fusion of epiphyses by contact (adhesions).
Serologic Features		
Sheep cell aglutination	Negative	Positive in 30% to 40% of the cases.

spinal signs In bedridden pain bound patients, in whom such an examination is not easy, the spinal manifestations may go unrecognized and only roentgenography will make the diagnosis possible Here it seems useful to present a table indicating the clinical differences between these two conditions, whatever may be their stage of evolution

DIAGNOSTIC PROBLEMS IN THE PHASE OF OBVIOUS DIAGNOSIS OR IN THE ADVANCED PHASE

These problems arise only in aged persons

Vertebral Osteo-arthritis

In the first place one may be easily misled by finding in roentgenograms, images that are manifestly osteo-arthritic and particularly osteophytes, and the diagnosis of Ankylosing Spondylitis is rejected here a systematic reading of films becomes essential one must carefully analyze the different spinal elements at different levels and thus it is possible to find a few typical syndesmophytes

Tabular Summary of the Differential Features of the Syndesmophyte and the Osteophyte at Their Beginning

(Figures 8 8bis and 9).

Bone Formation	*Syndesmophyte (bridging)*	*Osteophyte*
Direction	Vertical	Horizontal or oblique (it prolongs the vertebral plateau).
Aspect	Thin line or cottony appearance	Dense formation with sharp contour
Two Formations from a Single Pair of Vertebrae	Tendency to join, and they fuse when one reaches the other	Tendency to remain parallel.

But the contrary error must be avoided that is, to make too hastily a diagnosis of Ankylosing Spondylitis, which implies a graver prognosis and the necessity of repeated internal treatment Gold salts, Thorium X which may induce reactions and which

should not be employed in osteo-arthritis Indeed, some allow themselves to be influenced by certain clinical signs modification of the spinal curves, reduction in respiratory expansion, which are common in old men, and wrongly to regard certain bone formations as syndesmophytes the features of these formations must be carefully analyzed (Diagnosis of Syndesmophytes and Osteophytes, p 44)

In the same way certain conditions, such as the diffuse, decalcifying myelomatosis of Weissenbach and Lievre, Recklinghausen's bone disease, and the osteoporoses, also produce clinical symptoms of diffuse spinal disturbance it is also necessary to make a systematic study of the roentgenograms—a study which will reveal, besides osteoporosis, other roentgenographic spinal signs that are characteristic of Ankylosing Spondylitis and that, according to our experience, are always absent in cases of osteoporosis from other causes. We wish to emphasize the difficulty of interpreting the condition of the sacroiliac joints in roentgenograms of an osteoporotic pelvis.

Ankylosing Vertebral Hyperostosis of the Aged (81)

In old patients there exists an ankylosing disease of the spine, of which we have recently attempted to make an entity and which is distinguishable from Ankylosing Spondylitis by its clinical, roentgenographic, and anatomic features It seems legitimate to reproduce our description of this disease.

We have found in the literature anatomic descriptions of necropsy specimens which, to us, appear to be similar to those that we have studied in this disease. But no clinical and roentgenologic study of the subject has been published

In 1904 A Léri (136) gave an anatomic description of the spine of a patient who was afflicted with a disease which had been described by Pierre Marie and Astie under the designation (which can be criticized) of Bechterew's heredotraumatic kyphosis and which could well be applied to the kyphosis observed in our cases Moreover, Leri's old man presented an angulation of the spine of the Kümmel Verneuil type (post traumatic spondylitis), which had developed after a fall

M Meyer and E Forster (147) gave a similar anatomic descrip-

tion under the name of moniliform hyperostosis of the right side of the dorsal spine

In 1942 Oppenheimer (150) observed, *in old persons*, ossification of the spinal ligaments without involvement of the interapophyseal articulations These patients had a reasonable degree of spinal mobility and did not complain of pain He regarded these cases as belonging to the group of ossifications occurring in Ankylosing Spondylitis

In 1949 J Lacapere (131), in a study of spinal osteophytosis based solely on dry specimens, noted flowing ossified outgrowths which he called "melorheostose vertébrale", a term that may lead one to confuse this condition with the disease described under this name by A Léri in 1928 Except for this designation, Lacapère's anatomic description coincides, in its broad lines, with that of the previous writers and also with our own These flowing bony out growths *(coulees) were mentioned in the German literature by* Wenzell in 1824 (268) and by Rokitansky in 1856 (256) These authors, as well as later Schmorl and Junghanns (189), regarded these flowing outgrowths as osteophytes fused to a spinal column stiffened because of fibrosis of the discs

Our study of nine clinical and roentgenologic cases, of two roentgenologic records, and of two subjects that were dissected in the amphitheater, has enabled us to group certain clinical, anatomic, and roentgenologic features, and to isolate an entity from the group of ankylosing diseases of the spine.

Clinical Considerations

Age

This disease has been observed *only* in patients aged from 50 to 75 years (average, 65 years) This is in distinct opposition to Ankylosing Spondylitis *which is usually found in adolescent or* middle aged patients. Except in cases in which a definite ante cedent trauma *may have* played a part, the age of onset is difficult to determine because of the latent evolution of the disease

Sex

In our first communication we dealt only with men Since then ... feminine cases Ott (253) published one case

Symptoms

The patients present essentially an ankylosis of the dorsal segment, and often a marked stiffness of the lumbar segment In three of our cases (the 3 post traumatic cases) the stiffness extended to the cervical region The stiffness brought with it an awkwardness which was often moderate and well tolerated by the patients A few of them said they had become aware of the increasing stiffness, but in other cases the stiffness had been discovered on examination

In general the stiffness had become established without pain and, therefore, it was *latent* Only three of our cases had presented either a lumbago of short duration or sciatic pain At the time of examination the patients had practically no pain in the spine

We have not found any pronounced modification in spinal posture In two cases there was only a slight dorsal kyphosis of the type usually encountered in old men

Except in two cases in which the patients presented osteo-arthritis of the knees, which is a common condition at the age of these patients, the peripheral joints were not involved Therefore, there was no doubt that the disease affected only the spine

Given the advanced age of the patients, their general condition was satisfactory Six patients were notably obese while the weights of the others were normal

The rate of sedimentation was normal in all the cases, except one in which the patient was afflicted with chronic bronchitis In two patients the alkaline phosphatases were determined by the method of Bodanski, and they were 6 5 and 5 3 units or approximately normal

ROENTGENOLOGY

Lateral Roentgenography of the Spinal Column

It is this method of examination which permits one to make the diagnosis

Lateral roentgenograms of the dorsal spine furnish the most characteristic picture

In these can be seen a *continuous* but irregular, flowing band (coulée) of increased density which borders the anterior surface of the vertebral bodies and discs of the dorsal spine, and which extends to the lumbar region Its upper limit usually lies at about the

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fourth dorsal vertebra (Fig 118) in one case this band also covered the cervical spine *Opposite the vertebral bodies* its thickness varies from a *few millimeters to a centimeter*, and then the anteroposterior diameter of the vertebrae seems to be so increased that the vertebrae have a flattened appearance. More often the anterior border of the bodies appears distinctly visible at the posterior limit of the band Opposite the discs the band thickens into large blisters of a few centimeters The discs have a normal thickness and they do not project into these blisters the density of which is homogeneous (Figs 118 and 119)

Lateral View of the Lumbar Spine At the anterior surface of the column the flowing band is again seen, but here it is often discontinuous and broken opposite the discs or as if pushed back by the discs. Then may be seen curious and exuberant pictures in the form of a ' candle flame ', the tip of which may extend very high in front of the overlying vertebral body Here the anterior surface of the bodies forms a sharp line which permits one to distinguish it from the band (Figs. 120 and 121)

When the spinous processes are visible in the roentgenogram, they show an increased density of the supraspinous ligament, this is often very distinct in the posterior portion, where it may form a bridge from one spinous process to another

The structure of these bands is rather homogeneous and its texture is not apparent.

Lateral View of the Cervical Spine In the two cases in which we have been able to study roentgenograms of this segment, we have observed enormous bone formations on the anterior aspect of the vertebral bodies. These proliferations do not form a continuous band, but they are separated from one another by clear spaces in the projection of the disc spaces. These formations are very thick in the middle portion of the bodies which appear enlarged in the anteroposterior plane

This discontinuous appearance of the bony outgrowths is analogous to the appearance often seen in the lumbar region We believe that, in contrast to the continuity of the band in the dorsal region this discontinuity in the lumbar and cervical regions must be related to the greater mobility of these two regions

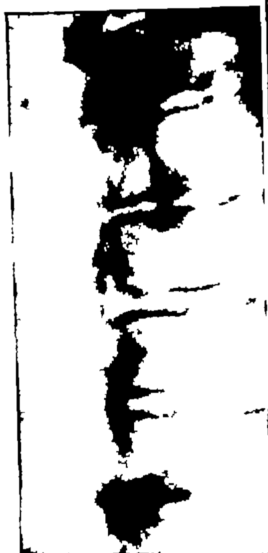


Figure 118



Figure 119

Figure 118 Mr Sil Lateral roentgenogram of the dorsal spine anterior flowing band of increased density beginning at D-4 This band covers the discs and vertebral bodies, but the anterior borders of the bodies can be distinguished.

Figure 119 Mr Per Lateral roentgenogram of the dorsal spine anterior band with large blisters opposite the discs the blisters do not project into the disc spaces.

Figure 120



Figure 121

Figure 120 Same patient as in Figure 118 Lateral roentgenogram of the lumbar spine picture of an anterior flowing band covering the discs and vertebral bodies opposite several discs the band is not continuous.

Figure 121 Same patient as in Figure 119 Lateral roentgenogram of the lumbar spine picture of hyperostosis in the form of a candle flame. Opposite L-3 and L-4 the base of the hyperostosis covers the entire anterior surface of the vertebral body



Figure 122 Same patient as in Figures 119 and 121 Antero-posterior roentgenogram of the lumbar spine and sacroiliac joints dense and fleecy bone structure of the third fourth and fifth lumbar vertebrae This appearance is due to hyperostosis of the anterior surface of the vertebrae (Fig 121) The sacroiliac joints are normal.

Antero-Posterior Roentgenography of the Dorsal and Lumbar Spine

The images are less characteristic and their interpretation is more difficult

Along the lateral borders of the spine the flowing outgrowths are hardly visible, but sometimes they may appear in the form of small bridges, especially on the right side. In a few cases, in the dorso-lumbar region, these bridges may be rather pronounced, and these images may resemble those of Ankylosing Spondylitis. Never theless, the contrast between the strong band visible in the anterior surface of the spine in lateral roentgenograms and the faintness of the lateral bands in antero-posterior roentgenograms is very striking, and this is one of the most important elements in the diagnosis. In Ankylosing Spondylitis, in fact, the syndesmophytes are much more visible in antero-posterior roentgenograms than in lateral roentgenograms

Another characteristic appearance, which is a result of the anterior position of the band, is "a fogginess of structure of the vertebral bodies" which is visible in antero-posterior roentgenograms and which is due to the superimposition of the band images over those of the vertebral bodies

The 'structure of the vertebral bodies" assumes a fleecy more or less dense and granular, aspect, and this image may lead to errors in diagnostic interpretation Paget's disease, neoplasmic process, etc A lateral roentgenogram reveals, in the anterior surface of the vertebral bodies, the superimposition of a band nearly 1 5 cm. thick which enables one to understand the appear ance of 'fogginess of structure" noted in antero-posterior roentgenograms (Fig 122)

Antero-Posterior Roentgenography of the Pelvis

The sacroiliac joints are visible and free from pathologic changes (Fig 122) In no case have we found any sign of fusion or any image of arthritis These pictures are in opposition to those of the bilateral sacroiliac lesions found in patients who are afflicted with Ankylosing Spondylitis. The iliac crests are irregular, their contours appear to be thickened sometimes throughout their length, backward from the posterior superior spine of the

ilium The increased density of this part of the bone extends over a third of the sacroiliac joint space and may give a false image of 'fusion "

Roentgenograms of the hands and extremities taken of patients in an attempt to determine changes in the periosteum, which might permit us to relate this disease to others that are better known and that have peripheral manifestations (pulmonary hypertrophic osteoarthropathy, mélorhéostose, etc), have given completely negative results

Differential Diagnosis

Vertebral Osteo-Arthritis

The question of differential diagnosis does not arise in the dorsal part of the spine, where the continuous flowing outgrowth, which is visible in lateral roentgenograms, is situated at the antero-lateral aspect of normal bodies and discs. This band differs from the usually small osteophytic beaks seen in osteoarthritis of the dorsal spine, in which the discs have become abnormally thin and the density of the vertebral plateaux has increased

In the lumbar region the continuous band can readily be distinguished from osteophytes, but when the outgrowths are independent of one another, in the form of flame like wisps, they may be confused with osteophytes. They differ from them by the following features (Fig 18)

They have a wide base that extends throughout or over a large part of the anterior surface of the vertebral body

The formation starts from the entire anterior surface of the vertebral body and a projection extends upward, there are never any large flame-like wisps starting from the lower border of the vertebra and extending downward

The thickness of the discs is usually normal.

The diagnosis is difficult because since hyperostosis develops in old people, osteo-arthritic lesions may be present in addition to hyperostosis. But in such cases lateral roentgenograms of the dorsal region show a continuous flowing band which establishes the diagnosis

Ankylosing Spondylitis

DIFFERENTIAL SIGNS BETWEEN ANKYLOSING SPONDYLITIS AND ANKYLOSING HYPEROSTOSIS

Ankylosing Spondylitis	*Ankylosing Hyperostosis*
Clinical	
In young or old men	In old men.
Kyphosis in 63% of the cases	Normal station of the spine
Lumbar and dorsal ankylosis, frequent involvement of the cervical region	Stiffness of the spine lumbar and dorsal rarely cervical.
Marked pain in the spine in nearly all cases	No pain in the spine.
Peripheral joint involvement in 72.5% of the cases	No involvement of peripheral joints.
Invalidism common	No functional impairment.
Increased sedimentation rate in 75% of the cases	*Normal sedimentation rate*
Roentgenography	
Syndesmophytes visible especially in antero-posterior films	*Visible band, especially in lateral films.*
Ossification present only in the upper and lower thirds of the vertebral body adjacent to the disc space	Thick band throughout the vertical thickness of the vertebral body
Sacroiliac changes	Normal sacroiliac joints.

ANATOMIC STUDY

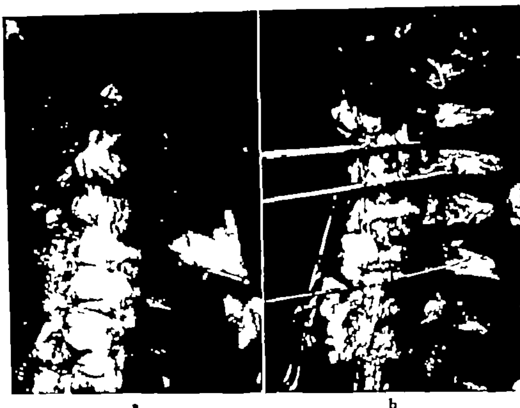
Our attention having been stimulated by our clinical and roentgenologic observations, we again took up the study of anatomo-roentgenologic records made by one of us before the war on the spines of old persons that had been obtained at necropsy

After study and comparison of the roentgenograms of these specimens with those of our clinical cases, we were able to consider two of them as belonging to the disease which we are considering and to bring together the anatomic, clinical and roentgenologic features. Here is the report of one of these necropsies

Specimen A

Lumbar mobility is complete in all directions, but the entire dorsal segment is absolutely rigid

Appearance of the Vertebral Column (Fig 123) A regularly wavy bony plaque is situated on the right side of the dorsal spine from D-2 to D-11 This plaque is continuous and embraces the hollows and bulges of the spine and it is crossed by transverse



a b

Figure 123 a. Photograph of Specimen A from the front bony plaque with blisters situated on the right side of the dorsal spine from D-3 to D-11 Noteworthy are the transverse furrows that mark the passage of blood vessels issuing from the aorta. b. Photograph of Specimen A from the left side the common anterior vertebral ligament is pushed back toward the left side by the bony plaque The tongue like, hyperostotic extensions of this plaque toward the left side are clearly visible. Opposite these the vertebral ligament can be easily detached.

furrows that mark the passage of vessels issuing from the aorta These vessels are seen in the fibrous sheath (around the spine) which had previously been reflected without difficulty with a periosteal elevator The bony plaque has the color and consistence of compact, pearly, white bone comparable with those of the femoral diaphysis. Here and there it is pierced by the nutrient vascular foramina Although this plaque is unilateral, it sends out tongue like prolongations at the level of the intervertebral spaces (D-7 and D-8, D-8 and D-9 and D-9 and D 10)

The Common Anterior Vertebral Ligament (Fig 123b) This is readily visible the bony plaque crowds it toward the left side, and the small tongues referred to are situated beneath the ligament

This ligament, which adheres strongly to the vertebral bodies as well as to the margins of the discs, can be easily loosened from the transverse tongue like projections that have already been described

The costovertebral articulations, the interapophyseal articulations, as well as the sacroiliac joints, have preserved their mobility

Roentgenogram of Specimen A Lateral View of the Dorsal Spine A well-defined flowing outgrowth extends in front of the bodies and discs from D-4 to D-12 It seems to hug the anterior surface of the vertebrae, and a clearer space is visible between the posterior part of the band and the *anterior surface of the vertebrae* (Fig 124)

Oblique View of the Dorso-lumbar Spine Here we find the band already described In this projection a sharp line separating the anterior surface of the vertebra from the bone band is not seen, but at certain levels it appears in this projection distinctly fused to the vertebral bodies its cancellous layer is continuous with that of the bodies (Fig 125) The contrast between the image of a band *hugging* the vertebral bodies, as seen in the lateral view and the image of a band *fused* to the vertebral bodies, as seen *in the right* oblique view, seems to be explained by the position of the hyper ostoses of the vertebral bodies In the present cases, indeed, the hyperostoses are situated in the right antero-lateral segment of the circumference of the vertebral bodies. *In lateral* roentgenograms there are two tangential surfaces 1) *that of the anterior surface of the body 2) that of the anterior part of the hyperostosis In the* oblique projection only one line is tangential to the beam of rays and, *since the hyperostosis is the most anterior* formation it will produce an image Thus, in this oblique projection, we see the seat of the hyperostosis, and roentgenograms clearly show that, in this plane, there is no line of cleavage between the hyperostosis and the vertebral body The cancellous layer of the hyperostosis and that of the vertebral body are continuous This explanation enables one to understand the appearance of ' juxtaposition of the hyper ostoses over the lateral images in our roentgenologic records.

Antero-posterior View of the Dorsal Spine A flowing out growth is seen to descend along the right side of the spine it forms bridges and, being superimposed over the vertebral bodies, it in creases the opacity of their right half



Figure 124



Figure 125

Figure 124 Necropsy specimen A. Lateral roentgenogram of the dorsal spine the anterior flowing outgrowth is situated in front of the vertebral bodies from which it appears to stand out distinctly

Figure 125 Same specimen as in Figure 124 Right oblique roentgenogram here the flowing outgrowth appears to be distinctly fused with the vertebral bodies.

In brief, from the point of view of pathologic anatomy, this disease is characterized by the presence of bony excrescences that arise along the entire sides of the vertebral bodies and pass in the form of bridges over the disc spaces, opposite which they often present offshoots. These hyperostoses have a bony structure with a very thin cortex. The layer of cancellous bone is continuous with that of the vertebral bodies

These proliferations of bone are usually situated in the dorsal spine, but in some cases they may extend farther from the axis down to the upper part of the sacrum. They tend chiefly to cover the right antero-lateral portion of the dorsal spine, but in the dorsolumbar and lumbar regions, they may also occupy the left antero-lateral aspect, and even the median anterior part, of the vertebral bodies

These proliferations of bone appear to be independent of the common anterior vertebral ligament contrary to the ligament, they are, in fact, lateral rather than median In our two anatomic examinations the ligament was normal and could readily be detached from the bony excrescences In one case in which the hyperostoses were situated not only on the right side of the spine, but also at its anterior part, they crowded the anterior vertebral ligament toward the left side

The large swellings or hyperostotic excrescences that often develop opposite the discs are not related to the salient margins of the discs We have a few cases of hyperostosis without any osteophytes being visible at other levels in roentgenograms When the spinal column presents large osteophytes, the bony proliferations cover the osteophytes and produce opposite them an additional prominence, but the largest proliferations are often seen opposite discs of normal thickness.

The co-existence of osteophytes and hyperostotic formations, which has just been described has often led physicians to interpret this condition as an advanced form of vertebral osteophytosis (osteo-arthritis of the spine) At the present time, from a roentgenographic point of view, we distinguish the osteophyte from the flowing outgrowth or band But only repeated anatomic examinations will permit one to say when a hyperostotic flowing band is or is not related to degeneration of a disc

Etiology

We do not have any definite ideas on the etiology of this disease One constant fact is the *senility* of the patients average age, 65 years Age, therefore, plays a role in the process

A severe trauma has been found three times at the beginning of the disease, we consider this as perhaps the precipitating cause of the clinical signs

The remaining difficulties presented by these patients must not be given too much weight these are people of advanced age who present the visceral sequelae of the collective diseases from which they have suffered during their entire life (cardiac insufficiency, bronchitis, icterus)

Consequently we believe we have the clinical, roentgenologic, and anatomo-pathologic elements necessary to permit us to isolate an ankylosing disease of the spine distinctly different from Ankylosing Spondylitis. For this disease we propose the designation of *senile ankylosing hyperostosis of the spinal column* this designation brings into one group 1) hyperostosis, which is the most obvious anatomo-pathologic element 2) and two constant clinical features stiffness of the spine and advanced age of the patient (average, 65 years) Doubtless this disease is not rare but most frequently it occurs without being recognized or it is confused with other ossifications of the spine.

We do not know the pathogenesis of this condition It should be noted that this flowing outgrowth tends to develop especially in the dorsal segment which, in old persons, already has little mobility According to some writers, this loss of mobility may permit the fusion of the osteophytes which usually remain independent

Therapeutic Deductions

Since the stiffness interferes very little with the activities of these patients, and moreover, since they are old people, the problem of treatment for the spinal disease rarely arises Attention is almost always attracted by other organic disorders of old age or by additional osteo-arthritic manifestations.

Etiology

As causes of Ankylosing Spondylitis the infections are most frequently mentioned, but none of the different hypotheses which have been advanced has been proved [1] Therefore, we will only mention the factors that may precipitate or favor the development of the disease without being its determining factors.

The infections gonorrhea, Malta fever, dysentery, staphylococcic or streptococcic infection, tonsillitis, pharyngitis, scarlet fever,

Tuberculosis
Icterus
Traumatism,
Cold, overwork, captivity
Pregnancy
Psoriasis
Heredity

GONORRHEA

In our patients its presence has been systematically sought, it was found among the patients antecedents in 31 out of 200 cases.

Gonorrhea had *immediately preceded the onset of the disease* in seven cases, or 3 5% *It is interesting to note that, in six of these cases, the first symptoms had been an attack of peripheral oligo-articular* or poly articular arthritis with a rather acute onset and that, in most of the cases, this attack had been followed by a long remission, the persistent involvement of the spine having supervened only after many years (5 to 10 years)

Example of Onset of Ankylosing Spondylitis with Gonorrhea

Mr Ch chemist aged 41 years, seen in October 1949

At the age of 25 years, he had contracted a very severe blennor

[1] The possible relationships between acute rheumatic fever rheumatoid arthritis, and Ankylosing Spondylitis are considered in the chapter on Classification, p. 285.

rhagia with fever, and this had lasted six months Eight days after the onset of the gonococcic infection pain and swelling of the ankles and knees very sharp pain in the hips and in a few joints of the hands The patient had remained in bed for six months after which he had resumed his normal work during a remission that had continued for three years.

At the age of 28 years, limping and transient pain in the right hip

At the age of 31 years, he was drafted into the army after having been stationed in a very humid trench, he had begun to feel pain in the hips and in the lumbosacral region A roentgenogram of the spine, which was made at that time, showed dorsolumbar syndesmophytes

Since then, the progression of the disease has been continuous now he is bedridden and an invalid

In this example gonorrhea has a distinct chronologic relationship to the onset of the disease which belongs to the febrile polyarticular type then a complete remission continued for three years After a period of overwork, the patient had another spinal attack a roentgenogram showed syndesmophytes, which proved that the disease had been progressing for a long time and that its onset had been synchronous with the articular attack at the age of 25 years

In the 18 cases in which an acute or chronic gonococcic infection had preceded by many years the onset of Ankylosing Spondylitis we have not observed a febrile onset which had been so frequent in the previous series a relationship between the two conditions cannot be proved

In six cases the gonorrhea had occurred *after the onset of the disease*, in two of these cases, after the blennorrhagia had abated, a fresh exacerbation of Ankylosing Spondylitis had occurred

Thus there are a few rather distinct cases in which the onset of the disease or a subsequent exacerbation is related to gonorrhea These facts have been considered in the study of involvement of the sacroiliac joints, a form of involvement which some regard as gonococcic but, to us, this idea seems to lack foundation

MALTA FEVER

We do not know a single case in which the disease has had its origin in Malta fever A report (95), which is often cited in the

literature, was based on five cases, but the author did not furnish any roentgenographic evidence and, according to his comments, it is difficult to know whether these cases were examples of Ankylosing Spondylitis or of melitococcic spondylitis

TONSILLITIS, PHARYNGITIS AND OTHER PRESUMABLY STREPTOCOCCIC INFECTIONS

These are rarely found at the outset of the disease, and we have found only three examples in our records in these cases a febrile polyarticular onset had been observed In two other cases these conditions had developed at the beginning of exacerbations in the course of the disease thus the part played by streptococcic infections in the onset or in exacerbations of the disease seems probable, but it does not occur frequently

STAPHYLOCOCCIC INFECTION

A general furunculosis that had progressed for six years, and a carbuncle that had lasted 45 days, had preceded the onset of the disease

SYPHILITIC INFECTION

Four of our patients were *syphilitics* but we do not have any reason to incriminate the treponema in the development of Ankylosing Spondylitis. This proportion of 2% is not very different from that which we have observed in our rheumatic patients as a whole and, considering the frequency of the disease we regard this as a mere coincidence

TUBERCULOSIS

In our patients we have not carried out any systematic investigation of the visceral manifestations of tuberculosis roentgenograms of the lungs were made only occasionally We cannot give any idea of the frequency of this condition in Ankylosing Spondylitis Many writers have emphasized the important or preponderant role

of Koch bacilli in this disease but, in our opinion, they lacked arguments and convincing statistics (179)

We have observed a few cases in which the *pulmonary disease had preceded or accompanied* the onset of Ankylosing Spondylitis (p 179), it is probable that tuberculosis had played a role in the development of Ankylosing Spondylitis, but no one could speak of tuberculous spondylitis because we have not found, in the joints, any tubercle bacilli or any lesions characteristic of tuberculosis, this is proved especially by the large number of histobacteriologic examinations that have been made, especially since the practice of arthroplasties began To us it would seem, rather, that tuberculosis might have a precipitating action that would modify the terrain

We have gathered two cases in which marked destruction of two adjacent vertebral bodies (Fig 37) is present this picture is similar to that of Pott s disease, but this diagnosis could not be proved According to Romanus and Yden, this destruction may be due to the inflammatory process of Ankylosing Spondylitis Usually, according to these authors, only slight erosions of the vertebral bodies are present—erosions which, in the few cases cited by them, seem to have preceded the appearance of the bony bridges in these cases destruction of the vertebral bodies was more marked

ICTERUS

Jaundice, whatever may be its origin, exhibits spectacular and transitory remissions, which Hench especially has emphasized for rheumatoid arthritis But also, and we have noted it in some cases, the first symptoms of the disease, or of exacerbations when the disease had been in a period of remission for a long time, may have appeared soon after the jaundice had disappeared, and this is probably due to a loss of resistance of the tissues the general fatigue that follows jaundice is a common phenomenon

TRAUMATISM

At the beginning of the disease we have obtained clear evidence of trauma in six cases, or in 5% of our cases.

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We have gathered two cases in which marked destruction of two adjacent vertebral bodies (Fig 37) is present, this picture is similar to that of Pott's disease, but this diagnosis could not be proved According to Romanus and Yden, this destruction may be due to the inflammatory process of Ankylosing Spondylitis Usually, according to these authors, only slight erosions of the vertebral bodies are present—erosions which, in the few cases cited by them, seem to have preceded the appearance of the bony bridges in these cases destruction of the vertebral bodies was more marked

ICTERUS

Jaundice, whatever may be its origin, exhibits spectacular and transitory remissions, which Hench especially has emphasized for rheumatoid arthritis. But also, and we have noted it in some cases, the first symptoms of the disease, or of exacerbations when the disease had been in a period of remission for a long time may have appeared soon after the jaundice had disappeared, and this is probably due to a loss of resistance of the tissues, the general fatigue that follows jaundice is a common phenomenon

TRAUMATISM

At the beginning of the disease we have obtained clear evidence of trauma in six cases, or in 5% of our cases

This very low figure, as compared with those given by other writers, is explained by the fact that we have deemed it wise to eliminate many cases in which the patients themselves had related the onset of their Ankylosing Spondylitis to a trauma, but in which *this etiologic notion did not stand* up under criticism for one or several of the following reasons

1) Close questioning disclosed clinical signs of the disease before the date of the trauma

2) Roentgenograms made *soon after* the accident, indicated lesions of long standing which could not have developed in a few weeks or months

3) Sometimes roentgenograms made before the trauma had revealed lesions typical of Ankylosing Spondylitis, especially in the sacroiliac joints, but which previously had not been identified as such.

Example of Ankylosing Spondylitis with Onset after Trauma

Mr Ker aged 52 years. June 16, 1937

At the age of 17 years, fell from a bicycle on the right hip after this fall pain in the posterior part of the right thigh some months later, pain in the posterior part of the left thigh, then pain in the *lumbar region* A *diagnosis of Pott's disease* was made and the patient was sent to Leysin where he was treated for 18 months At the end of his stay there, the error was corrected and the diagnosis of Ankylosing Spondylitis was made

At the age of 30 years, iritis appeared and this ocular disturbance alternated from one eye to the other and recurred almost every year At the present time the patient continues to suffer from his spine.

On examination, besides complete stiffness of the spine, he presents slight involvement of the right shoulder the sedimentation rate varies from 28 to 57

This example shows that trauma may play a precipitating role in the onset of a few cases of Ankylosing Spondylitis

Our cases have not presented features that would permit us to group them into a distinctive clinical form progression of the disease in the spine has been identical to that of ordinary Ankylosing Spondylitis involvement of peripheral joints has occurred with the same frequency as in cases without a traumatic onset

Sometimes trauma has precipitated a fresh attack in a patient who had been afflicted with Ankylosing Spondylitis for many years

FATIGUES OF WAR, COLD, OVERWORK

One of these factors played a part in nine of our cases, these were in men who had been prisoners of war (8 cases) or who had fought in the war (1 case)

In six of these cases the clinical onset had been noted during their captivity, and in three others an exacerbation had supervened in the course of these physical trials (Example, Chat , p 246)

PREGNANCY

In two of our cases the disease had begun after confinement (in one case immediately afterwards, and in the other 6 months afterwards) In a third case an exacerbation had followed the patient's confinement In a fourth case, in which marked stiffness of both hips had already developed, recourse to a Cesarian section had to be made

In one case the disease had begun after hysterectomy, in another patient an exacerbation and aggravation of the disease had occurred after the same operation From such meager evidence conclusions cannot be drawn

PSORIASIS

We have collected 13 cases in which psoriasis had been associated with Ankylosing Spondylitis, they constituted a clinical form and they were mentioned previously (p 199) But if this dermatosis gives to Ankylosing Spondylitis a special impress, it does not appear that it can be regarded as an etiologic factor

DYSENTERY

We have not observed Reiter's syndrome in any of our cases

HEREDITARY FACTOR

We have found in our records 10 cases with familial antecedents parents, brothers or sisters who had been afflicted with Ankylosing Spondylitis On two occasions we have examined two cases in the same family, in which the patients did not present more common features than two cases of Ankylosing Spondylitis taken at random

This already ancient idea of familial Ankylosing Spondylitis, although it has been supported by few cases, has suggested that an hereditary factor might have an important or essential place in the etiology of Ankylosing Spondylitis But in order to study it, it would be necessary to examine clinically and roentgenologically the parents of each patient and not to limit one's self to simple information obtained by questioning, we do not have any definite ideas on the possible importance of heredity as an etiologic factor

During the past few years, different writers have carried out systematic studies of families one of whose members had been afflicted with Ankylosing Spondylitis

In 1936 Clausen and Kober (37) examined the families of 10 patients who were victims of Ankylosing Spondylitis, and in these 10 families they found nine cases of the disease. These patients were distributed as follows three cases in one family two cases in one family, and one case in each of four other families.

In 1948 B Rogoff and R. H Freyberg (173) examined the families of 24 patients, these families being suspect because of information furnished by the patients The authors admitted that, out of 100 parents of patients affected with Ankylosing Spondylitis, the percentage of those affected was from 9% to 13%

In 1949 H F West (218) after having calculated the frequency of the disease (1 in 2000 inhabitants of the city of Bristol [400,000 inhabitants]), estimated the frequency of the disease as 100 times greater in families one of whose members was being treated for Ankylosing Spondylitis (9 cases of Ankylosing Spondylitis in the families of 83 patients) than in the other families among this population

These studies have been confirmed by Stecher (206) who, after having examined a series of parents of patients afflicted with Ankylosing Spondylitis and a group of persons taken at random, found

that Ankylosing Spondylitis was 70 times more common among the parents of patients suffering from Ankylosing Spondylitis than in the control series

Moreover, Stecher noted that, of 10 women who were afflicted with Ankylosing Spondylitis and who had brothers, the brothers of four of these women were also affected by the disease

Stecher (206,264) thinks that "The disease is inherited as a single chromosome dominant factor with incomplete penetrance (pene trance the ratio of actually compared to expected affected) Penetrance differs greatly between the sexes, being 73% in men and 19% in women Spondylitis is not a sex linked character Penetrance in the family where there is a woman with a spondylitis is 100%."

Summarizing, different conditions, such as gonococcal infection, tuberculosis trauma overwork, and surgical operations, may coincide with the onset of Ankylosing Spondylitis or of its exacerbations. We regard these conditions, which are diverse and not specific, as precipitating factors and not as specific causes of the disease.

On the contrary, it is evident that there exists a dominant hereditary factor According to Stecher this factor is more penetrating in men than in women To us the cases in which precipitating factors are or are not found and the cases in which heredity is evident (familial cases) do not seem to differ from one another

Pathologic Anatomy

Our present knowledge concerning the pathologic anatomy of Ankylosing Spondylitis is based on a small number of necropsies, on a few studies of dry bones and especially on roentgenograms. The histo-pathologic data are still meager all the more so since most of the cases examined were ancient ones, and an accurate clinical history was lacking Only a greater number of necropsies on cases in which the disease has not been active a long time, and the removal of surgical specimens from patients who have been thoroughly studied from a clinical point of view, can make possible the solution of certain problems

History The first study of dry specimens was made by Connor in 1700 (43) The first necropsy seems to have been performed by Hilton Fagge in 1874 (57), but it was Pierre Marie and Léri who in 1906 (114), gave a complete macroscopic description of cases examined at necropsy, but which had been thoroughly studied clinically

We will now give

1) A personal description based on a study of 23 dry specimens of the spine, a few of which included the three spinal segments as well as the pelvis

2) The necropsy reports which, by furnishing information on the condition of the ligaments and intervertebral discs, complete our study of the dry specimens

3) A histologic sketch illustrated with sections made in Professor Rutishauser's service by G Riotton and P Wettstein and with comments by ourselves.

STUDY OF DRY SPECIMENS

Lumbar Region

This study is based on seven dry specimens.

The Vertebral Bodies

The shape of the vertebral bodies is not altered, and their height is normal

From a structural point of view, the external appearance of the vertebral bodies in the zones where they are not covered with syndesmophytes is normal The vascular foramina are quite visible (Figs 126a and b)

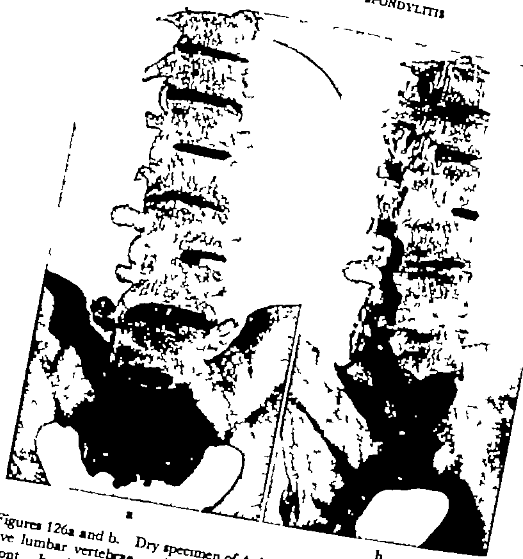
In a sagittal section of the spine the cancellous bone of a vertebra, and especially its central portion, sometimes appears to be rarefied, the trabeculae are fine and their number relatively small and they may even be lacking

This rarefaction of the cancellous bone is readily visible in roentgenograms, and especially in lateral roentgenograms (Fig 127) this generally appears in the form of clear zones which are arranged in horizontal bands that occupy the middle third of the vertebral body the Haversian and Henle canals are often very apparent. These pictures of rarefaction contrast with the generally denser images of osteo-arthritis of the spine

The Bone Bridges or Syndesmophytes, Their Sites and Insertions

The appearance of the vertebral bodies is modified by the presence of bone formations when these are *in a stage of moderate development* they are inserted into the upper or lower third of a vertebra, and in the form of bridges they cross the intervertebral space, where they present a swelling or bulge and insert themselves into the lower or upper third of the adjacent vertebra

These formations are not evenly distributed over the entire circumference of the body they are very large over their lateral and antero-lateral aspects, while they are thin or absent over the anterior aspect Thus the appearance of the lateral aspects of the lumbar spine is greatly modified opposite the intervertebral discs



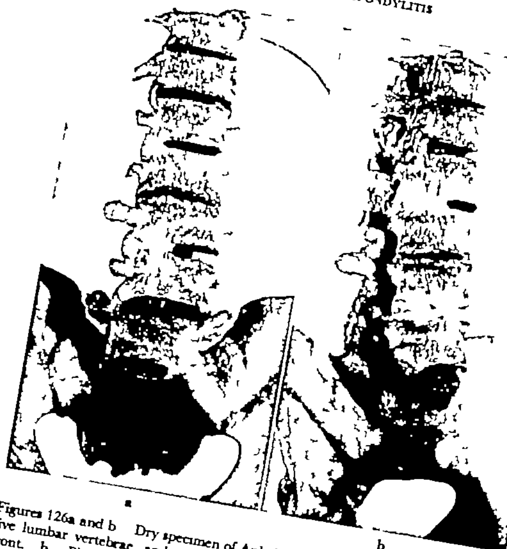
Figures 126a and b. Dry specimen of Ankylosing Spondylitis, including a pelvis, five lumbar vertebrae, and two dorsal vertebrae. a. Photographed from the front. b Photographed at a right three-quarter angle. (Specimen of J. Forestier R. Colbez, and P Robert)

and the adjacent parts of the vertebral bodies they present convexities and opposite the middle third of these bodies they present concavities, while the appearance of the anterior surface of the spine is either normal or only slightly altered

A transverse section of a specimen of Ankylosing Spondylitis (Fig 7c) shows that the dimensions of the vertebral plateau are normal the plateau is limited by the internal cortical layer of the syndesmophyte and is situated at the outer limit of the intervertebral disc A section of spine affected with osteo-arthritis, on the

[illegible]

[illegible]



Figures 126a and b Dry specimen of Ankylosing Spondylitis, including a pelvis, five lumbar vertebrae, and two dorsal vertebrae a. Photographed from the front. b Photographed at a right three-quarter angle. (Specimen of J Forestier R. Coltez, and P Robert)

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Figures 126c and d. Same specimen as in Figures 126a and b c. Roentgenogram at a right three-quarter angle d. Antero-posterior roentgenogram.

contrary, shows a widening of the vertebral plateau which is extended by the osteophyte

In a few specimens that presented marked alterations the bone formations also covered the middle third of the vertebral bodies, and this was especially true of their lateral aspects When this occurs, the syndesmophytes then form a continuous line, with bulges and hollows and these bulges and hollows persist because the syndesmophytes are always thinner opposite the middle third of the bodies. Even at this very advanced stage, the bone formations do not extend beyond the postero-lateral angle of the body, and they are absent from the posterior part of the bodies and discs

The bony outgrowths and bridges are whiter and smoother than the vertebral bodies, and they contain fewer vascular foramina

(Figs. 126a and b) Perhaps this difference in appearance is related to the osteoporosis affecting the bodies

A frontal section of the lumbar spine enables one to study the shape and structure of the bone bridge, as well as its relations to the disc and vertebral bodies (Fig 6a) This section reveals a *cortical layer called superficial* it is continuous and its thickness varies slightly opposite the middle third of the bridges it is very thin, but it is slightly thicker (1 mm.) opposite the middle third of the vertebral body, where it fuses with the old cortical layer of the vertebra Beneath the cortical layer is a layer of cancellous bone which is usually continuous with that of the vertebral body the cortical layer of the body having disappeared opposite the abut ments of the bony bridge sometimes vestiges of this old cortical layer may persist. Opposite the disc the cancellous layer of the bridge is limited in its deep portion by a *cortical layer called deep* which separates it from the disc

In a sagittal section of the same specimen we find in the anterior part of the spine and opposite the disc, a thin sheet of bone which, starting at right angles from the marginal border of the vertebra extends downward or vertically upward toward the opposite sheet (Fig 6b) At different levels all degrees of evolution of these anterior formations are found from those a few millimeters thick to those that form a complete sheet covering the intervertebral space this sheet does not bulge outwardly It should be recalled that these bone formations are very often absent from the anterior part of the spine

The Intervertebral Discs

In five specimens in which the lumbar segment was complete the disc maintained its normal thickness, except in one specimen (Fig 126a) in which the thickness of the disc between L-2 and L-3 was distinctly diminished this reduction in thickness affected the entire disc and was regular

Ossification of the internal substance of the disc was not found in a single specimen In the dry specimens the deep cortical layer appeared to be situated at the limit of the disc ring histologic sections (p 272) show that the syndesmophyte has its seat in the short fibers of the ligamentary system or in the space between these

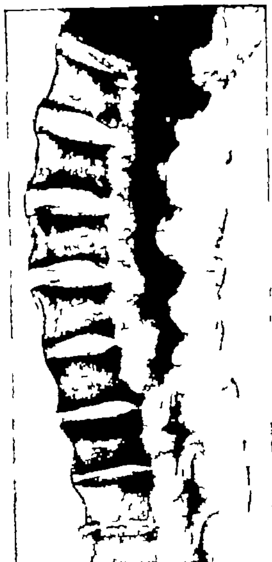


Figure 127 Dry specimen of Ankylosing Spondylitis Lateral roentgenogram of the lumbodorsal spine thin anterior syndesmophytes discs of normal thickness osteoporosis of the bodies, especially in their middle portion ankylosis of the interapophyseal joints ossification of the interspinous ligaments D-11 and D-12.

fibers and the outermost fibers of the disc At a more advanced stage the process of ossification also affects the latter fibers

The Interapophyseal Joints

These joints present a bony ankylosis and form, with the articular epiphyses, a small continuous column [1] *In a horizontal section* is found either a continuous layer of cancellous bone without limit, in which the vestiges of an old articular space can be recognized or more rarely a more or less long cleft situated at the site of the former

[1] In one specimen the volume of the interapophyseal joint had increased, as if the extremities of the bone or the articular capsule had proliferated.

articular space. *Ossification of the joint, therefore, begins at the periphery*

We consider entirely justified the remark made by Güntz (98) that the changes in the soft parts of the interapophyseal joints that are seen in fresh specimens may disappear during maceration of the dry specimen The observation, in a dry specimen, of apparently normal interapophyseal joints should not lead one to conclude that the specimen is not one of Ankylosing Spondylitis

The Ligamenta Flava

In place of the ligamenta flava there is found a sheet of bone with vertical striations, the thickness of which may vary between 1 and 2 mm In a few cases these bony sheets completely block the posterior interlaminar space

The Intervertebral Foramina

In the lumbar region the intervertebral foramina appear to be normal, they are bounded in front by the bodies and the discs which, opposite the postero-lateral angles of the vertebrae, do not present any prominence or ossification

The syndesmophytes that cover the lateral surface of the spinal column behind stop at a certain distance from the intervertebral foramina in a few cases, at a more advanced stage, we have seen syndesmophytes extending as far as the postero-lateral angles of the vertebrae but they did not encroach on the foramina

The posterior part of the foramen is limited behind by the ankylosed interapophyseal articulations, the increased size of which does not appear notably to reduce the caliber of the foramen Nevertheless, the contiguity of these altered articulations to the nerves running through the foramen may explain the radiating pain of the inflammatory phase that produces ankylosis

Only the intervertebral foramina between L-5 and S-1 and between D-12 and L-1 present some peculiarities which we will study

The Spinous Processes

Opposite their posterior border and sometimes also at their upper and lower borders, these processes present a bony thickening In one case we found in place of the supraspinous and interspinous

ligaments, a continuous sheet of bone situated between the spinous processes, this sheet extended forward and joined the ligamenta flava which were also ossified

The transverse processes are normal

Dorsal Region

We have studied 12 dry specimens of the dorsal spine (half the specimens included 12 vertebrae) of patients who had been afflicted with Ankylosing Spondylitis

Most of these dorsal spines presented a marked dorsal kyphosis

The Vertebral Bodies

Their structure presented the same alterations as in the lumbar spine, but the bones had undergone even greater rarefaction, frequently the layer of cancellous bone had disappeared

In roentgenograms the unsectioned specimen showed, in this segment, clear bands in the middle third of the vertebral bodies, and the canals of Henle were quite visible.

In two specimens we found, in the zone where the kyphosis had started, one or two wedge-shaped vertebral bodies in which rarefaction of bone was very pronounced In these very advanced cases the ankylosed spinal column presented a tubular appearance comparable with that of a long bone.

The Syndesmophytes

These have the same appearance as in the lumbar region, but their distribution is more extensive In this zone, indeed, are found prominent syndesmophytes that form real pads, not only on the lateral surface, but also on the anterior surface, in specimens with an aggravated kyphosis these syndesmophytes on the anterior surface are very marked

In two specimens, on the left side of the spine, beginning at D-6 there appeared a vertical impression that deviated slightly in its lower part *In this zone the syndesmophytic pads* were flattened and more compact, and they presented a smooth surface Their impression appeared to be related to the thoracic aorta

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The posterior part of the foramen is limited behind by the ankylosed interapophyseal articulations, the increased size of which does not appear notably to reduce the caliber of the foramen Nevertheless, the contiguity of these altered articulations to the nerves running through the foramen may explain the radiating pain of the inflammatory phase that produces ankylosis.

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In two specimens, on the left side of the spine, beginning at D 6, there appeared a vertical impression that deviated slightly in its lower part. In this zone the syndesmophytic pads were flattened and more compact, and they presented a smooth surface Their impression appeared to be related to the thoracic aorta

Often the entire thickness of the discs had diminished and they were narrowed in front, especially in the zone where kyphosis had begun The discs presented two kinds of ossification

Anterior Fusion of the Vertebral Bodies This was found in cases in which the anterior narrowing had brought into contact the two bony plateaux that had ended by fusing together, the cancellous layer of the vertebral bodies extended without any break in continuity

In these specimens the disc space no longer reached the anterior border of the spinal column which was thus transformed into a continuous bone, with transverse clefts, residues of the discs, and opening behind toward the spinal canal

Ossification of the Interior of the Discs In certain specimens in which the discs had preserved a reasonable thickness (Fig 128), we found in the discs small and more or less extensive zones of ossification with a cancellous type of structure, these ossifications contrasted with those previously described, in which the discs had become narrower

The Interapophyseal Articulations

Only in half the specimens had these undergone any alterations In other dry specimens, where these joints were intact, syndesmophytes were present in the dorsal region while in the cervical and lumbar segments of the same specimens changes in the interapophyseal joints were present.[1]

The Interspinous and Supraspinous Ligaments

Here one finds an ossification of the supraspinous ligament, which extends in the form of a bridge from one process to another In the dorsal region we have not found any ossification of the interspinous ligaments.

The Costo-Vertebral and Costo-Transverse Articulations

We have noted an ossification of the ligamentary apparatus that unites the head the rib the vertebra, and the costal tuberosity to the transverse process, and sometimes also [illegible] fication of the

[1] In serial roentgenogram [illegible] t seen changes [illegible] pophyseal joints, and then syndesmophytes, [illegible]

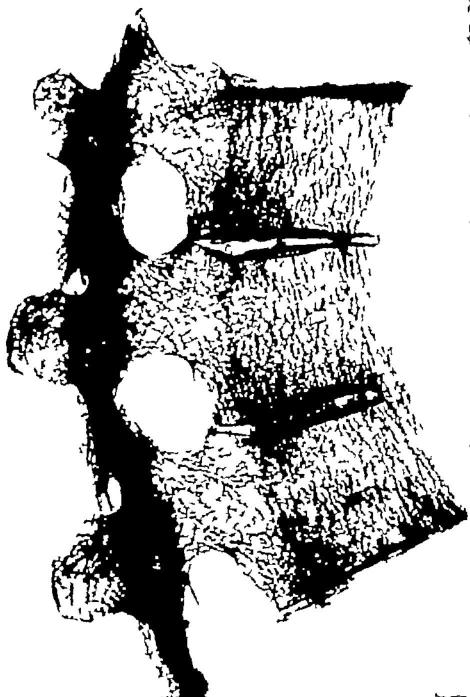


Figure 128 Dry specimen of Ankylosing Spondylitis. Lateral roentgenogram of the dorsal spine ossification of the anterior part of the discs.

cervico-transverse intercostal ligaments. Often enough, when the rib is separated from its insertion, the central zone of the costovertebral and costotransverse articular surfaces appears to be intact On the posterior surface of the transverse process we have found rather frequently some of the vertical striations that were described by Borellini (24)

In this region the intervertebral foramina are difficult to examine because of the posterior extremities of the ribs that have remained in place and because of the general ossification of the ligaments, which thus form narrow clefts through which run the intercostal nerves. This was noted from the intervertebral foramen between D-4 and D-5 as far as the foramen between D-11 and D 12, where the cleft between the ossified formations is very narrow

Cervical Region

We have examined six dry specimens

The Vertebral Bodies

They did not present any deformities, and in the sections their structure appeared to be normal

The Syndesmophytes

a In two dry specimens (Figs 129a, b c, d, and e) they resembled those in the lumbar region

b In three specimens (Fig 130a b and c), that present interapophyseal changes typical of Ankylosing Spondylitis, the appearance of the syndesmophytes is altogether peculiar, in fact, they appear in the form of a longitudinal plaque applied to the anterior surface of the vertebral bodies and discs the posterior surface of this plaque adheres to the vertebral bodies, and the anterior surface is flat and smooth as if it had been polished with sandpaper it laps over the lateral surfaces of the spine where it is much thinner The upper limit of this mass is at about C-3

Sagittal sections of these three specimens show that it is bone tissue with a superficial cortical layer, a deep cortical layer opposite the discs, and between the two a cancellous layer continuous with the cancellous bone of the vertebral bodies. This formation,

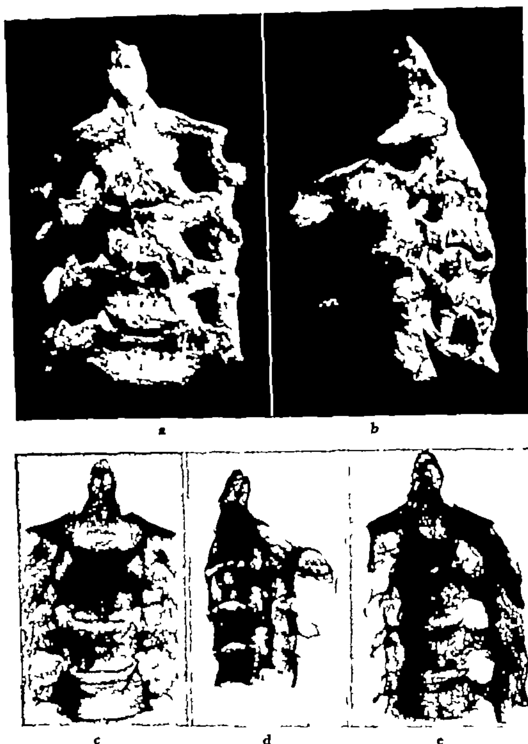


Figure 129 Dry specimen of Ankylosing Spondylitis. Cervical Spine a. Photographed from the front anterior syndesmophytes. b Photographed from the side bony ankylosis of the interapophyseal joints. c. Antero-posterior roentgenogram. d. Lateral roentgenogram. e. Roentgenogram at a three-quarter angle.

therefore, is identical to the typical syndesmophyte but here the syndesmophyte is much larger, especially in the middle portion of the vertebral bodies

This laminated zone resembles the zone we have seen on the left side of the spinal column, that is related to the aorta the organ in contact with the spine is the esophagus which, conceivably by its *movements of deglutition, shapes the syndesmophyte*

The Interapophyseal Articulations

These joints present changes that are identical to those observed in the lumbar region The ankylosed joints and the articular processes form elegant little columns.

The Uncovertebral Articulations

They are fused in nearly all cases, transverse sections show an ankylosis analogous to the ankylosis described in relation to the interapophyseal articulations

The Intervertebral Foramina

These are quite free the foramina through which passes the vertebral artery are not altered

THE SACRO-ILIAC JOINTS

In two dry specimens the lesions are much less pronounced than one could have imagined from the roentgenograms the bones are not completely ankylosed and the articular space is abnormally thin, but between the two articular surfaces small clefts about 1 mm thick persist.

Again we find bone bridges between the small wing of the sacrum and the wing of the ilium the iliotransverse ligament is ossified

Intervertebral Foramen between L-5 and S-1

Normally this foramen is narrower and longer than the other foramina in the lumbar region indeed, it forms a true tunnel (30)

But in our specimens this foramen does not show any pathologic reduction in caliber nevertheless, the presence of ossification in

a b


c

Figure 130 Dry specimen of Ankylosing Spondylitis. Cervical spine with changes of a peculiar type anterior bony plaque (Fig 44) a. Photographed from the front. b Photographed from the right side. c. Left lateral roentgenogram

the superior iliotransverse and iliosacral ligaments and of the syndesmophytes 'fills' the region

The Pelvis

The iliac crests (2 specimens) were thick, and in one case they presented a thick edge from which hung bony spines that could be compared with stalactites. On the outer surface of the pelvis rugosities and small spines were situated along the muscular lines

In one case the descending rami of the ischia were slightly larger than normal and roughened, especially at their lower border, we found, inserted into the ischial tuberosities and at the sites of the great sacro-sciatic ligaments, strong bony spines 4 centimeters long the ascending rami were thickened and rough, but the cleft of the pubic symphysis was preserved

The Hips

In three specimens the acetabulum was free from the head of the femur It was deeper than normal because the margin of the acetabulum extended outward in the form of a rough, circular excrescence (ossification of the acetabular rim) the ischio-pubic notch was slightly narrowed by proliferations that were inserted into the anterior and posterior horns, the bottom of the acetabulum was invaded, in its peripheral portion, by bony tissue from the semi lunar facet.

In a fourth dry specimen the head of the femur and the acetabulum were ankylosed Judging from a roentgenogram, the contours of the head and acetabulum had disappeared, the cancellous bone of the head and acetabulum was continuous from one to the other

NECROPSIES

Necropsies have shown that *the anterior longitudinal ligament of the spine is free* and independent of the syndesmophytes that lie immediately beneath it (137) (55), only in cases with very advanced disease did we find slight ossification of this ligament. The

surgical findings of J J Herbert, in the course of vertebral osteot omies (106), confirm the fact that the anterior longitudinal ligament is free, in order to reach the syndesmophytes this ligament must be retracted Histologic studies (89, 98, 41), which we will present later, confirm these findings We can, therefore, assert that, in Ankylosing Spondylitis, the anterior longitudinal ligament is not involved in the ossifying process

But some clarification of the definition of the anterior longitudinal ligament should be given

Classically and according to Poirier and Charpy, "the common anterior vertebral ligament is a long fibrous band or ribbon which extends over the middle portion of the anterior surface of the verte bral bodies from the basal apophysis of the occipital bone to the sacrum. The fibrous fasciculi that compose the anterior vertebral ligament extend longitudinally and run parallel to one another The long, superficial fasciculi extend over the bodies of several vertebrae (4 or 5), while the deep fasciculi, which are shorter, extend from one vertebra to the next, and they blend with the periosteum When the ligaments are separated by traction, which is possible, the periosteum of the vertebral bodies, which is intimately blended with the ligament, is torn at the same time, it is found that this adhesion or fusion, which is very strong opposite the vertebrae, is weak over the discs" (159)

Fr J Rainer (163) gives a different description, or more pre cisely a different definition, than the one given by Poirier and Charpy According to this author, the column formed by the vertebral bodies and discs would be surrounded by a fibrous sheath that unites the vertebrae one to another This fibrous sheath is composed of 1) The external elements of the disc, or fibrous ring, and a sheath composed of short fibers that are inserted into two adjacent vertebrae and thus pass in the form of a bridge over the corresponding intervertebral space This fibrous coat, which Rainer calls *peri rachis*", completely surrounds the lateral and anterior aspects of the vertebral bodies 2) A system of long super ficial fibers that form the common anterior vertebral ligament These fibers are inserted not like the preceding ones into adjacent vertebrae, but into vertebrae that are remote one from the other This ligament forms a sort of band which, in the lumbar region

covers only the anterior surfaces of the vertebral bodies The deeper fibers of this ligament are shorter and shorter and form an imperceptible transition with the short fibers of the 'peri rachis.'

We believe the *ossifications* that were described in our study of the dry specimens develop chiefly at the expense of what Rainer describes as "perirachis disc ring and system of short fibers, which are continuous with the periosteum of the vertebral bodies Later the ossifying process may affect the middle fibers of the fibrous coat, and very rarely the anterior longitudinal ligament.

This explains how it is possible to separate the common anterior vertebral ligament, or the system of long fibers, from the short fibers which, as anatomists have found, are ossified Syndesmophytes predominate over the lateral aspects of the spine, where the common anterior vertebral ligament is not present [1] the ossifications, therefore, develop at the expense of the short fibers that cover the lateral aspects as well as the anterior surface

Histologic Studies

The histologic studies (199 82, 136, 98, 89, 41, 269 265, 228, and 258) were based on a small number of cases. Rarely did they include the spine and the joints of the extremities of the same patient which would have made it possible to compare the lesions by the affected regions or joints

The reported cases may be divided into two groups according to the presence or absence of inflammatory signs But, as Freund (89) aptly remarked, different authors have not the same criteria to define a rheumatic inflammatory lesion For him the lesions found in certain cases of Ankylosing Spondylitis and those found in rheumatoid arthritis are not identical. Nevertheless, Freund believes that the presence or absence of inflammatory signs permits one to divide the cases of Ankylosing Spondylitis into two groups.

We will analyze the different histologic studies and present the information placed at our disposal by Professor Rutishauser (269, 258)

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[1] Perhaps the anterior longitudinal ligament is responsible for the slight development of anterior syndesmophytes, because it exerts pressure on them.

The histologic sections were obtained from the spine of a patient aged 41 years, who had died of chronic nephritis In his case the Ankylosing Spondylitis appears to have started at the age of 17 years with a polyarticular episode It was not until very late that attention was drawn to the kyphotic deformity of the spine and that a roentgenogram of the spine revealed the existence of Ankylosing Spondylitis (latent form) But in spite of the long standing disease, some of the changes in certain regions had not progressed much

SPINE

The Disc Changes, and the Bony Bridges Opposite and Near the Discs

Not a single writer has found any signs of inflammation in the anterior part of the spine. Most of the papers relate to the ossification of the outer part of the disc which ends in the formation of a bony bridge opposite the disc.

Ossification of the Disc We have drawn especially from the writings of Rutishauser and his co-workers, and from those of Van Swaay

"The fibrous ring loses its fibro-cartilaginous character and assumes a hyalin aspect. Then, around the cells of the interstitial substance, it gradually presents a calcareous impregnation This impregnation begins in the region adjacent to the marginal border and extends vertically towards the corresponding formation in the vicinity of the adjacent border This impregnation foretells and directs the ossification which follows it. It is a question of *metaplasia* of the disc ring in bony tissue This bone undergoes a change in structure and becomes transformed into cancellous bone with its cavities continuous with those of the vertebral body

The formation is vertical and it occupies the place of the outer most fibers of the disc ring The anterior longitudinal ligament is not involved in this transformation therefore, it is not affected by the ossifying process

As this bone bridge progresses toward the center of the disc, it may become thicker, in very advanced cases the entire disc, even its posterior portion, may become ossified For a long time the

cartilaginous plaques remain free from this change Signs of inflammation are not found at any stage (Rutishauser, 269)

Sections of Bone Bridges at their Outset, Their Situation *Disc between D-4 and D-5* At this level roentgenograms show a normal image By contrast a sagittal section through this area discloses marked histologic changes.

The main lesion is found in the superficial layers of the fibrous ring of the disc Its central portion shows large foci of mucoid degeneration with rare areas of regeneration beginning in the fibro-cartilage of the disc A little deeper than the cortical layer *of the vertebra is found a sharply circumscribed cavity about 1 mm* in diameter, containing very large cells some of which resemble fibrocytes while other parts are made up of cells with small, deeply stained nuclei surrounded by a clear halo, the cytoplasm is very abundant and contains granulations that are faintly stained with eosin It is possible to follow the formation of these cells which derive from the fibro-cartilage seeking to repair the necrosis.

In the sections that we have examined we have not seen any vessels The intercellular substance contains some edema fluid with very few fibrils

Between this lesion and the anterior ligament there is another pathologic focus, with ill-defined contours, and with massive necrosis and mucoid degeneration of the fibro-cartilage In other sections through the same disc the two foci are fused (Fig 131a)

Similar lesions are not found in the anterior ligament, but a few small foci of adipose tissue undergoing necrosis are seen between the cortical layer of the vertebra and the anterior ligament.

The great excess of calcium extending over 2 or 3 mm. in the anterior part of the terminal cartilaginous laminae of the vertebral bodies should be emphasized The fine calcareous granulations are in large measure pericellular In this particular zone, however, they infiltrate the interstitial substance and accumulate along a line with geographic contours opposite the posterior margin

Disc between D-5 and D-6 (Fig 131b) In this para-sagittal section we find, directly beneath the insertion fasciculi of the psoas muscle, a thin lamina of bone crossed by a few *Haversian* canals. Above and below it rests on the cortical layer of the verte bra Between this bone and the fibro-cartilage of the disc are seen

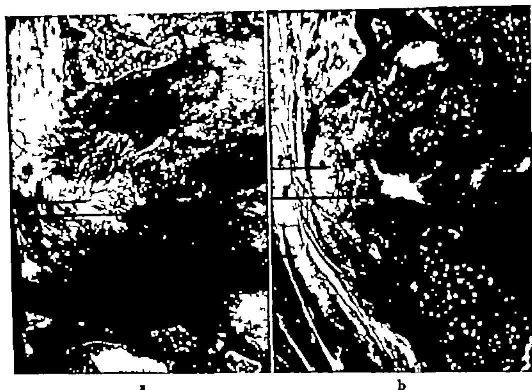


Figure 131 a. Disc between D-4 and D-5 (3× magnification) Sagittal section. Stain, hematoxylin and eosin 1 posterior part of the anterior longitudinal ligament 2. focus of mucoid degeneration and necrosis opposite the fibrous ring 3 zone of fibrocytic proliferation with beginning regeneration 4 massive impregnation with calcium opposite the terminal lamina of cartilage. b Disc between D-5 and D-6 (3× magnification) Para-sagittal section. Stain, hematoxylin and eosin 1 lateral portion of the anterior ligament and muscular fibers of the psoas muscle 2. syndesmophytes replacing the fibrous ring 3 residue of the degenerated and necrosed fibrous ring small islands of calcification 4 disc proper (the gap between the necrosed fibers and the anterior part of the disc is an artefact)

a few areas of connective tissue derived from the fibrous ring, these are largely necrosed The orientation of these residual fibers is in large measure modified, and a few of them are impregnated with calcium and stain deeply with hematoxylin

The same observations are to be made in the posterior portion, but there ossification is less advanced

Opposite the disc proper only degenerative changes are to be seen, and these should be related to the age of the patient

These two images, the earliest we have ever encountered, prove that the *initial lesions occur in the fibrous ring of the disc and not in the*

ligament They would seem to begin by mucoid degeneration of the fibers of the ring and to be accompanied by an excessive deposition of calcium at the level of the marginal border

Section of Bone Bridge in an Advanced Stage of Development. Disc between D-6 and D 7 Sagittal section In this area a roentgenogram shows a thin bridge of bone connecting the anterior borders of the two vertebrae and corresponding to the classical description of the syndesmophyte.

Here we observe first that the anterior vertebral ligament is made up of parallel fibers stained a rose color by eosin and not taking hematoxylin nowhere is there any trace of calcium deposit This ligament presents the same features as the anterior vertebral ligament of a healthy spine taken from a patient of the same age, which we have examined histologically But instead of resting against the fibrous ring of the disc, it covers a compact band of bone which is continuous with the cortical layers of the vertebral bodies above and below it, this sheet of bone is in direct contact, in its posterior part, with the *intervertebral disc which is very slightly altered but no longer* has a fibrous ring At the osteo-cartilaginous limit, which stains more deeply with hematoxylin a few small vascular buds are to be seen (Fig 132)

Sections Showing the Extension of the Lesions Toward the Center of the Disc. Disc between D-7 and D-8 Sagittal section Here we find the same arrangement as in the previous section, but the anterior bridge of bone is thicker It is the seat of a rearrangement of bone shown by the enlargement of the canals into cavities containing bone marrow and by the modified orientation of the suture lines which are no longer parallel to one another Besides, *the ossification* follows the penetration of the vascular buds into the interior of the disc (Fig 133)

Disc between D-8 and D 9 Sagittal section The anterior limit of bone is formed by a thin, compact lamina, the remainder of the bone bridge being formed by areolar bone in continuity with that of the vertebral bodies it contains a mixed bone marrow The anterior part of the disc is slightly altered, but in its median portion there appears hyalin cartilage in contact with which is trabecular bone rich in mixed bone marrow Worthy of note is the normal thickness of the disc and the persistence, opposite the ossification of



Figure 132



Figure 133

Figure 132. Sagitta section of a syndesmophyte (D-6–D-7) Stained with hematoxylin and eosin. (5 X magnification) 1 lamellar ossification of the fibrous ring of the disc 2. intact anterior vertebral ligament (the separation at the top is artificial, as is also the multiple split appearance of the disc).

Figure 133 Sagittal section of the disc between D-7 and D-8 1 Penetration of the vessels into the disc, leading to ossification 2. Rearrangement of the lamellar bone which gradually becomes areolar 3 Intact anterior vertebral ligament.

the median zone, of two bands of cartilage (upper and lower) these are nearly straight and they indicate the limits of the disc in process of ossification (Fig 134)

Disc between L-5 and S-1 Sagittal section Two thin bands of hyalin cartilage mark the limits of the transformed disc. The anterior portion is formed by cancellous bone with rather dense trabeculae Behind, the bridge of bone is thinner, but it has the same features as the anterior part. In the center of the greatly transformed disc is noticed a very large area of fatty marrow measuring 12 X 6 mm in diameter bordered in part by incurving trabeculae and in part by the residual cartilaginous bands. This



Figure 134 Sagittal section of the disc between D-8 and D-9 showing beginning ossification of the latter The syndesmophyte is composed of cancellous bone

fatty tissue is undergoing massive necrosis, and this is accompanied by the appearance of fibrinous deposits opposite the walls of the necrosed fat cells. In the anterior part as well as in the posterior part the ligamental fasciculi are found intact (Fig 135)

Comments[1]

In the discs we find nearly all the stages of their transformation, from the lamellar ossification of the fibrous ring only to ossification of the entire disc

We wish to draw attention to the fact that this ossification at the outer border of the disc corresponds, from the roentgenologic point of view, to an image like a bony bridge which practically does not project beyond the line of the vertebral bodies.

Moreover, we have observed that, in these cases, the vertebral bodies have a square shape To Romanus and Yden (257) this

[1] The case studied by Riotton and Wettstein was one with a latent evolution the roentgenogram is similar to that shown in Figure 27 In this latent form the disc changes appear much more diffuse and pronounced than in the other forms in which the disc has preserved its normal thickness.

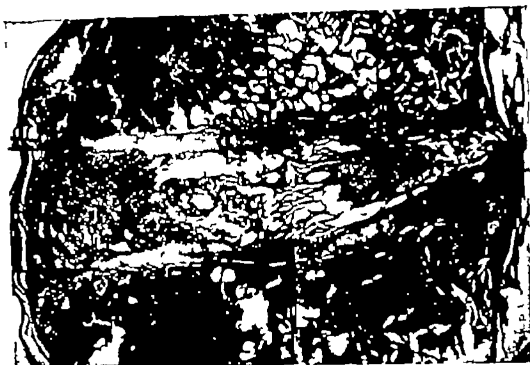


Figure 135 Sagittal section through the disc between L-5 and S-1 Hematoxylin-eosin stain (Magnification 3X) 1 Anterior vertebral ligament pushed back by the bone bridge 2 Cartilage residues corresponding to the upper and lower limits of the disc 3 Active foci of enchondral ossification 4 Partial ossification of the fibrous ring in the posterior part of the disc 5 Large focus of necrosis in the midst of the medullary adipose tissue, with deposits of fibrin (to be noted is the absence of bone throughout this entire zone)

would seem to be due to a destruction of the vertebral angles by an inflammatory process, we are not familiar with the histologic data of these authors.

Ossification adjacent to the Disc. There is another roentgenographic image (Figures 8, 8bis, 9, 10, 12, and 14) in which the ossification begins in the upper or lower third of the lateral aspect of the vertebral body We have found this type of bone bridge in some of our dry specimens. Freund has shown histologic sections in which the bony outgrowth had developed beneath the perichondrium or the perispinal periosteum. The ossification may extend either inward toward the disc or outward and very rarely toward the median fibers and the longitudinal ligament

Changes in the Interapophyseal Joints As we have previously mentioned, the reports may be divided into two groups according to the presence or absence of inflammatory signs.

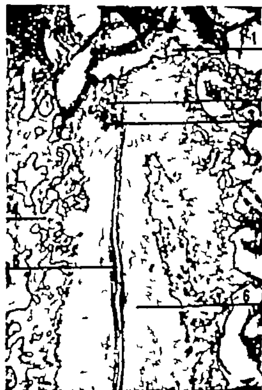


Figure 136 Vertical section through the posterior articulation between L-1 and L-2. Hematoxylin-eosin stain. (Magnification 3×) 1 Inner limit of the ossified capsule 2 Progression of ankylosis by enchondral ossification (no trace of inflammation) 3 Preparatory perichondrocytic calcification 4 Subchondral osteosclerosis 5 Mucoid degeneration of the cartilage 6 Zone of multiplication of the cartilage cells.

Ankylosis by Enchondral Ossification The majority of joints examined by Riotton and Wettstein had already become ossified therefore the onset and the ossification of the capsules could not be observed The ankylosis progresses centripetally from the joint capsule, where it begins, toward the center of the joint This is a classic enchondral ossification without any trace of inflammation

Sections of Interapophyseal Joints Not a single articulation is absolutely normal A vertical section through the arc D-2 and D-3 shows, at one point of the inferior apophysis, a few myelogenous buds penetrating the cartilage and accompanied by a few round cells of lymphocytic type The cartilage is relatively well preserved The residues of the superior diarthrosis, limited at each extremity by trabeculated bone that has invaded both capsule and cartilage, no longer show any articular cleft

In a vertical section at the level of L-1 and L-2 (Fig 136) different kinds of changes in the cartilages are observed The upper part of the joint is invaded by cancellous bone with mixed marrow which, beginning at the outer margin of the now ossified joint



Figure 137 Horizontal section through a posterior articulation between D-10 and D-11 Hematoxylin and eosin stain (Magnification 3×) Complete bony ankylosis. 1 Ossified articular capsule 2. Normal ligamentum flavum.



Figure 138 Central part of the previous section (Magnification 10×) Cartilaginous rests in process of ossification

capsule, stops abruptly against the residual cartilages Here many changes are seen There are large areas in which the cartilage cells have proliferated and one may see as many as 12 cartilage cells piled one against another and surrounded by a diffuse, bluish halo At other points, especially at the two ends of the subchondral zone, fine deposits of calcium surround the cartilage cells Small myelogenous buds extend toward these areas some are even bordered by bands of a rosy substance corresponding to osteoid The articular cleft persists and opposite the cartila

ginous surfaces, there is mucoid degeneration with exposure of the fibers beneath, may be seen wide bands in which the cells have disappeared The subchondral bone, which is distinctly more dense than normal, sends a few rare myelogenous buds in the direction of the cartilage

A horizontal section through the vertebral articulations between D 10 and D-11 shows a marked bony ankylosis. In this area there are still a few islands of cartilage impregnated with fine granules that stain deeply with hematoxylin and that correspond to calcareous deposits which nearly always surround groups of cartilage cells. These cartilaginous rests, which occupy the center of the former articulation, are here and there eroded by small, vascular, myelogenous buds that do not contain any inflammatory cells At certain points one can recognize the former articulation only by the orientation of the thickened bone trabeculae elsewhere every trace of the joint has disappeared, and the trabeculae extend without any break in continuity from one articular process to another (Figs. 137 and 138)

A vertical section through the articulation between D-11 and D-12 shows still more advanced bony ankylosis without any areas of cartilage

In the other interapophyseal articulations that we examined we found the same images as those which have just been described

Van Swaay (265) found a similar process of ossification He stressed the point that the obliteration of the articular cleft is due to proliferation of the two cartilages which invade each other and thus results in a cartilaginous ankylosis Subsequently, according to him, an enchondral type of ossification develops.

Fibrous Ankylosis Following an Arthritis Siven, Fraenkel, Güntz, and Freund found inflammatory lesions lymphocytic nodules, pannus beginning in the capsule and gradually destroying the cartilage The three first authors regarded these lesions as identical to those of rheumatoid arthritis, but Freund does not share this opinion

Güntz, and Freund, make a distinction between the inflammatory lesions in the interapophyseal joints and the enchondral ossification of the disc According to them the latter is due to the loss of spinal mobility resulting from ankylosis of the interapophyseal joints.

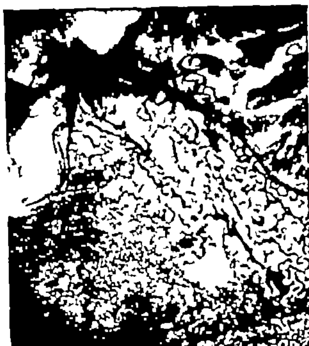


Figure 139 Frontal section through the sacroiliac region Complete bony ankylosis. Only rare islands of cartilage remain



Figure 140 Section through the hip of a woman, aged 53 years with Ankylosing Spondylitis. Destruction of the non-degenerated cartilage by the inflammatory pannus (Section furnished by Prof E Rutishauser)



Figure 141 Section through the hip of a patient aged 25 years, with Ankylosing Spondylitis. Lymphocytic infiltration with a few plasma cells in a pannus covering and absorbing the articular cartilage of the femoral head (Section furnished by Prof E. Rutishauser)

Changes in the Costovertebral and Costotransverse Joints Wettstein and Riotton have observed ankylosis by enchondral ossification Siven, on the contrary, observed destruction of cartilage by a destructive process

CHANGES IN THE MANUBRIO-STERNAL JOINTS Savill studied five manubriosternal joints he did not find any signs of inflammation but he thought the replacement fibrous tissue which he described was similar to that found in advanced rheumatoid arthritis

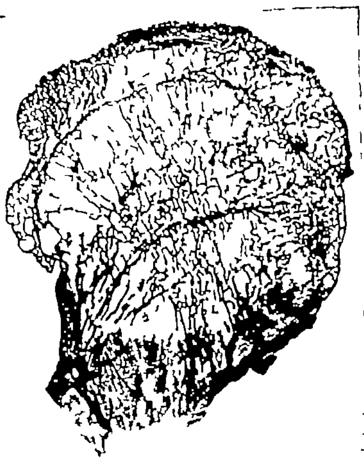


Figure 142. Section through the hip of a man aged 29 years, with Ankylosing Spondylitis. In this section the contour of the femoral head, as well as that of the acetabulum, are only slightly altered. The articular cartilages are replaced by porotic bone in continuity with that of the acetabulum (small, close meshed trabeculae in the upper part of the section) and of the head. In the supero-internal quadrants of the old articular space small bands of fibrous tissue persist. In the head the bone is porotic and oncotic. Hematopoiesis begins only at the level of the femoral neck. (Section furnished by Prof E. Rutishauser)

Sacro-iliac Joints

Van Swaay while studying a case with recent lesions, found, in the sacroiliac joints, the same type of enchondral ossification as in the interapophyseal joints

Joints of the Extremities

The hips have been most frequently studied because of their more frequent involvement on the one hand and, on the other hand, because of the tissue removed in the course of arthroplasties

Most writers found inflammatory signs characterized by a pannus that destroyed the cartilage Cruickshank (228) found this in the 13 cases which he studied, and Rutishauser (258) in three out of four cases The histologic lesions were identical to those found in rheumatoid arthritis Nevertheless, except in one case, the destructions observed in our roentgenograms are much less pronounced than in rheumatoid arthritis and they do not change the shape of the femoral head

The fourth case studied by Rutishauser showed ossification of the cartilage in this case the articular space had preserved its normal width, as was shown by the vestiges of the joint surfaces As we have already mentioned, (p 133), we often find a roentgenographic image of bony ankylosis with an articular space of normal width, this picture corresponds to this type of lesion Unfortunately, we do not have any histologic sections made during the initial stages of this bony ankylosis

Summarizing, in Ankylosing Spondylitis two kinds of processes have been described 1) An enchondral ossification featured at the outset by calcareous deposits around the cartilage cells, without any trace of inflammation This kind of ossification has been observed in the intervertebral discs, the interapophyseal joints, and the sacroiliac joints To us it seems that the bony ankylosis of a hip studied by Rutishauser must have been a result of this process 2) An inflammatory process with formation of a pannus that destroys the cartilage and may end in fibrous ankylosis This has been observed in the joints, and especially in the hips As far as the hips are concerned, the inflammatory lesions are similar to those of rheumatoid arthritis.

At the present time it is impossible to say whether or not these different types of processes are associated in the same patient if they should be, their character would then depend on their situation Neither can it be claimed that there are two anatomic forms of Ankylosing Spondylitis.

Classification

In the chapter on Etiology we mentioned that the cause of Ankylosing Spondylitis is unknown Different hypotheses have been proposed, but these lack substantial foundations In this chapter we will first present the clinical and anatomic features peculiar to Ankylosing Spondylitis, then we will set forth the relationships between Ankylosing Spondylitis and the other inflammatory rheumatic syndromes.

Comments on the Characteristic Features of Ankylosing Spondylitis

If an hereditary factor is admitted, the part which it plays in the disease is not understood From the analysis of the symptoms and from their development certain ideas emerge which help to understand the disease

Clinical Features The sacroiliac joints become involved early and constantly Nearly always involvement of the spine extends upward Peripheral involvement develops in an unregulated manner, and this is in contrast to ascending involvement of the spine Peripheral involvement affects especially the joints of the hands and feet, and those of the lower extremities

Anatomic Features Ankylosing Spondylitis involves essentially formations derived from connective tissue the locomotor apparatus and the iris. It may end in ankylosis by ossification of the joint elements capsule, cartilage, fibro-cartilage (disc), and tendon insertion

In the joints and discs this ossification develops centripetally, beginning at the periphery it may reach the center of the articular cleft or of the disc

The presence or absence of inflammatory signs observed in early

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In the joints and discs this ossification develops centripetally, beginning at the periphery, it may reach the center of the articular cleft or of the disc

The presence or absence of inflammatory signs observed in early

lesions makes the rare histologic reports contradictory This contradiction may be only apparent, and in this event different kinds of anatomic changes may occur

Biologic Features. Until now biologic research has yielded only negative data absence of streptococcic antibodies which are present in cases of acute rheumatic fever, absence of streptococcic agglutination phenomena and of agglutination of sensitized sheep cells which, in contrast, are present in a high percentage of cases of rheumatoid arthritis. Acid and alkaline phosphatases and blood calcium are at normal levels Only changes in sedimentation rate and in proteins are found, but these are not specific.

This ossifying process, which most frequently develops in young men, may continue throughout their lives The cause of this process is unknown The hypotheses advanced to explain the initial involvement of the sacroiliac joints (J Forestier) have not been proved, the same is true of the part played by the lymphatics (J Forestier) or by the Venous System (Batson) in trying to explain the ascending progression of certain spinal conditions

RELATIONSHIPS BETWEEN ANKYLOSING SPONDYLITIS AND OTHER FORMS OF INFLAMMATORY RHEUMATISM

At the present time acute rheumatic fever rheumatoid arthritis, and Ankylosing Spondylitis are only clinical syndromes, some writers, basing their claims on the differences between these three syndromes, regard them as distinct conditions, while others contend that they are difficult to distinguish and that they are really one and the same disease that can produce a variety of clinical pictures We will now present the arguments of these different writers

ANKYLOSING SPONDYLITIS AND ACUTE RHEUMATIC FEVER (BOUILLAUD'S DISEASE, RHEUMATIC FEVER)

The idea of relationship between these two clinical entities is based essentially on the observation, in patients afflicted with Ankylosing Spondylitis, of symptoms which, in principle, are characteristic of acute rheumatic fever, that is

1) Transitory attacks of articular inflammation at the beginning of the disease,

2) Cardiac lesions

Transitory Attacks of Articular Inflammation

At the beginning of the disease, these attacks have been recorded by different writers (35, 54) and especially by Fischer and Wuntz, who found them in 14% of their cases of Ankylosing Spondylitis (58) We have observed these symptoms or noted them in the histories of 8 5% of our cases (Onset of the Disease, p 178)

In our opinion, although the articular attacks are similar to those of acute rheumatic fever, they differ from them by

1) The pain and swelling remain confined to the joints that are affected at the beginning

2) The frequent involvement of the joints of the fingers and toes, and of the cervical spine,

3) The tendency of the disease to persist in one or more articulations.

It is obvious that these are only clinical shadings and that differentiation between an early attack of Ankylosing Spondylitis and one of acute rheumatic fever may be impossible, but we do not believe this to be a decisive argument in favor of the identity of the two processes In fact, a polyarticular syndrome with swelling and transitory pain may be observed in infectious diseases, such as Malta fever and streptococcic infection Nothing therefore, justifies one in classifying any transient attack of articular inflammation as acute rheumatic fever

Cardiac Manifestations

During these past few years, anglo-saxon writers have studied the cardiac lesions of "rheumatoid arthritis' from a clinical and anatomo-pathologic point of view (177, 14, 171)

Their reports indicate that 1) in "rheumatoid arthritis" the frequency of clinical involvement of the heart would seem to be low or rare (171) 3 4%, that is, about the same proportion as that found in a series of control cases (2%) 2) anatomic lesions of the heart are found in a high percentage of cases of "rheumatoid

arthritis" (177) The writers do not agree on the point of knowing whether these anatomic lesions are similar to those of acute rheumatic fever or whether they are clearly different (Bennett, 14) In these studies the cases of Ankylosing Spondylitis were not separated from those of rheumatoid arthritis.

In Ankylosing Spondylitis Krebs and Wurms (125) often found involvement of the myocardium or endocardium this involvement would seem to be more common in cases with an acute polyarticular onset. Bernstein and Broch (17) while reviewing 352 cases of Ankylosing Spondylitis, found clinical involvement of the heart in 16 cases or in 4 3% of the entire series these included aortic insufficiency with or without stenosis (6 cases), mitral stenosis (4 cases) heart block (2 cases) pericarditis (2 cases), and auricular fibrillation (2 cases) In these 16 cases polyarticular involvement had been present at the outset of the disease Out of 190 cases in which electrocardiograms had been made, abnormalities had been found in 47 cases, and 62% of them had had disturbances of conduction (increased P-Q interval) The peripheral rheumatic syndrome had been found twice as often in patients with abnormal electrocardiographic findings.

In brief, 11 9% of all cases had either clinical signs or electrocardiographic deviations The authors realized that the acute peripheral rheumatic syndrome plays a very important part in the etiology of cardiac symptoms in Ankylosing Spondylitis (17) In this disease the cardiac manifestations are benign, even in cases with valvular defects, even in aged persons, the signs of myocardial insufficiency are not very pronounced

In our patients electrocardiographic examinations were not made, but working in the environment of a spa with hot springs, we have been led to a careful clinical examination of the circulatory system before subjecting them to treatment.

In our 200 cases of Ankylosing Spondylitis we have found but three cases of cardiac involvement two of these had mitral stenosis in the third case it was impossible to determine the type of lesion present, but among the patient's antecedents chorea was present. In all three cases the involvement was benign it was always well compensated and did not raise any therapeutic problems. These patients tolerated the spa treatment without fatigue All three

patients had had transient attacks of articular inflammation at the beginning of the disease

Concerning the origin of these cardiac lesions several hypotheses may be considered 1) Ankylosing Spondylitis itself may produce cardiac lesions 2) Ankylosing Spondylitis and acute rheumatic fever are but different manifestations of one and the same disease, 3) in one patient the presence of Ankylosing Spondylitis and acute rheumatic fever with cardiac disturbance was a mere coincidence

Present knowledge does not yet make it possible to solve the problem of the possible relationship between acute rheumatic fever and Ankylosing Spondylitis, but *from a practical point of view*, several facts should be considered

The clinical features, the evolution, and the treatment of the two diseases are completely different In a case of Ankylosing Spondylitis, even with cardiopathy, the classical treatment of acute rheumatic fever (treatment with salicylates) does not appear to be indicated the treatment is the same as for the arthritides in general,

In Ankylosing Spondylitis the cardiopathies seem to be benign and do not raise any particular therapeutic problems A reservation must be made that patients with severe cardiopathies should preferably be referred to a cardiologist so we have had an opportunity to observe only benign cases

It is wrong to consider the polyarticular attacks at the onset of Ankylosing Spondylitis as a symptom of acute rheumatic fever, in our opinion, these are simply acute and transient exacerbations of a type of chronic inflammatory rheumatism

ARE ANKYLOSING SPONDYLITIS AND RHEUMATOID ARTHRITIS TWO VARIETIES OF THE SAME DISEASE, OR ARE THEY DIFFERENT CLINICAL ENTITIES?

In favor of the first idea the following arguments may be cited

Peripheral articular localizations of inflammatory type are present in a very large proportion of cases of Ankylosing Spondylitis.

The constitutional and general symptoms, as well as the increase in the sedimentation rate are observed equally in the two diseases

The pathologic changes that develop in the interapophyseal articulations during the active period of the disease resemble those found in the peripheral joints in rheumatoid arthritis.

These arguments have seduced American writers and have caused them to accept the idea of a single disease hence the name "Rheumatoid Spondylitis' which has been given to Ankylosing Spondylitis

On the contrary, we believe—and this is the opinion of the majority of French and, we think, of European writers—that *the two diseases are really two different clinical entities* (Table, p 229) and that they should not be confused Here are some arguments in favor of this opinion

The distribution of the disease in the two sexes is inverted since Ankylosing Spondylitis is more common among men, while rheumatoid arthritis is more common among women,

Ankylosing Spondylitis never begins during early infancy or in old age, rheumatoid arthritis may develop in infants and in old persons,

Peripheral involvement in the course of Ankylosing Spondylitis has a sporadic, oligo-articular distribution it almost never causes dislocation and, as far as the large joints are concerned, it leads to bony ankyloses in the most extensive forms (the panarthritic form) bony ankylosis reaches its maximum In rheumatoid arthritis, on the contrary peripheral involvement is systematic, symmetrical, dislocating and causes deviation, but it rarely ends in clinical ankyloses of the large joints. In rheumatoid arthritis the lumbar, dorsal or thoracic signs are not found (in this disease we have noted only 20% of cervical involvement),

Meynet's fibrous nodules are never observed even in advanced cases of Ankylosing Spondylitis, while they were found in 15% of our cases of rheumatoid arthritis

Especially in Spondylitis, ankylosis by ossification of the articular elements is common (ankylosis of the sacroiliacs, syndesmophytes, ankylosis of the hips), while it does not occur in rheumatoid arthritis in which, on the contrary, stiffness by contact of the ulcerated bony surfaces and more or less marked osteolytic phenomena are nearly constant

The inflammatory muscular nodules (88, 90), which consist of pericapillary collections of lymphocytes alone or associated with plasma cells, epithelioid cells, and sometimes eosinophiles, seem to be characteristic of rheumatoid arthritis (207), because they have not been found in patients who are afflicted with Ankylosing Spondylitis

In conclusion, we should regard the three great rheumatic syndromes as different clinical entities Laboratory studies may perhaps confirm these clinical data

Treatment

Ankylosing Spondylitis is a *chronic disease* over many years it progresses (study of cases with more than 20 years' evolution, p 187) usually in spurts separated by more or less long periods of spontaneous stabilization Because of this *the physician should when dealing with a patient who is afflicted with Ankylosing Spondylitis, follow him regularly all his life*

A close clinical study, according to a scheme that has already been described (p 16) and repeated every six months or every year, is essential, it should be completed by a *biologic examination* of the blood (rate of sedimentation) and, when necessary, by a *roentgenographic check* which should bear chiefly on the lumbodorsal region and, whenever necessary on certain joints such as the hips and knees

The purpose of this observation is, essentially *to determine disturbances in spinal posture* that is, kyphosis, and *faulty postures of the articulations of the lower extremities*, and *everlastingly to repeat to the patient the rules of orthopedic hygiene* that allow the inflamed joints to rest in good position, that is, in the position of function

But this observation does not necessarily imply continual therapeutic medication Physicians should have the same attitude as specialists in syphilis and diabetes who have acquired the notion of chronic disease and the way to treat it. Chemotherapeutic procedures, radioactive or hormonal agents should be used with discrimination because they have rather definite indications according to the phases of the disease, the signs of its progress, and also its localizing signs.

We will divide the treatment of Ankylosing Spondylitis in the following manner

1) Rules of postural and orthopedic hygiene to prevent defective postures and deformities.

2) Basic treatment treatment with roentgen rays and medicines to which should be added treatment with general radioactive agents by injection

3) Heat treatment.

4) Corrective orthopedics

5) Surgery

Then we will present a plan of treatment to deal with different eventualities

RULES OF POSTURAL AND ORTHOPEDIC HYGIENE TO PREVENT DEFORMITIES

Except in circumstances that will be mentioned later or except in a severe attack of progressive disease, there is no value in recommending rest in bed The patients have every advantage in carrying on moderate physical activity, which should be regulated as much as possible for each patient, this should include short active periods interrupted by frequent periods of rest

In Ankylosing Spondylitis a strict rule should be for the patient to take his rest in the *recumbent position* During these rest periods, care should be taken to put the affected joints in physiologic positions the disappearance of the inflammatory phenomena will thus be favored, and stiffness in a faulty position will be avoided in a patient who has merely been advised to remain in bed during a period of active disease, the physician will be greatly surprised when the patient resumes the standing position, to note ankyloses in a defective position, which themselves will cause disability Rest in bed, therefore, should be carried out under certain conditions

The resting position of the joints is the position of function for the spine and the large joints of the extremities we will indicate the defective postures that may develop and that the physician must combat.

The Spine The patient tends to become round shouldered when he goes to bed, therefore, he should lie in the dorsal position and the bed should not be too soft and should not become hollow in the center To avoid this a hard wooden board should be placed between the box spring and the mattress This board should be large enough to cover the entire length of the box spring and the width of the trunk (hard bed minimum 1 20 m. by 0 60 m) The

patient's head should rest on a thin pillow Thus the spine remains straight and, if stiffening should develop, it will occur with the trunk and the neck in a correct position

The Hips and Knees Even when the patient is lying down, these joints tend to assume a flexed position This is often encouraged by the patient's family or by attendants who, in order to relieve him, slide pillows under the knees The physician should strictly forbid the use of such pillows

The Ankles and Feet. On the one hand, the weight of the bed coverings leads to foot-drop in equine position with flexion deformity of the toes (Fig 113), and to a small degree of valgus and external rotation of the lower extremity, which constitute one of the most serious obstacles to a resumption of walking This may be avoided by eliminating the weight of the bed coverings by means of a hoop and by keeping the feet in good position by means of sand bags placed under the soles of the feet and against the outer part of the feet

It is easy to understand how *prolonged rest in an easy chair should favor the development of defective postures* With the patient's hips and knees in flexion he allows himself to lean forward, that is, he "rounds" his back, flattens his thorax, and lowers his abdominal organs. It is therefore justifiable to call the easy chair ' the enemy of the rheumatic In Ankylosing Spondylitis rest should be taken on a sofa or chaise longue, and the sitting position should be reduced to a minimum

Full confinement to bed is indicated only in cases in which the disease progresses rapidly in spite of treatment, in those that present marked involvement of the lower extremities, and in those in which the disease is accompanied by marked impairment of the general condition fever loss of weight, pallor, excessive perspiration and tachycardia.

In these bedridden patients, besides precautions concerning posture, the following measures should be taken 1) cautious daily mobilization of the affected joints, especially active mobilization 2) take care of the condition of the skin in fact, the peripheral circulation of these patients is impaired and the skin is dry with a minimum of subcutaneous adipose tissue Bedridden patients should be washed daily and they should be lightly rubbed with

camphorated alcohol and kept warm in order to favor the circulation Their rooms should be light and well ventilated, and if possible should be oriented toward the sun [1]

When, at certain periods of the disease, rest in bed is indispensable, its prolongation entails certain undesirable effects anorexia, constipation, muscular atrophy, and a tendency to stiffening For these reasons the patient should not be required to stay in bed any longer than is necessary He should be kept in bed as long as severe inflammation of the joints of the lower extremities or a markedly defective posture of the hips and knees persists

Resumption of the standing position and of walking should be done very gradually, at first for a few minutes, and then for a few hours This period of rehabilitation may continue for many months or even for many years During this period the patient is frequently tempted to rest in an easy chair for many hours our experience has convinced us that, as a general rule, no patient should be allowed to rest in a sitting position for more than one hour, if he works in a sitting position, he should, from time to time, rise, attempt to straighten as much as possible, and take a few steps. It should not be forgotten that we have observed more frequent and more marked defective postures of the lower extremities in sedentary patients who, for instance, worked at a desk than in patients who had done hard physical labor

When the patient resumes his professional or household activities, he should take, as long as the disease shows signs of progression, ten or twelve hours sleep at night, and his day should be broken by rest in the horizontal position for at least half an hour a day

Gutter Splints or Plaster Shells (210, 211, 68, 73)

Simple rest in bed is not sufficient when marked inflammatory involvement with a tendency to defective posture is present. At the beginning this is not always manifest clinically but, at the time of periodic examinations, a systematic search for it should be made

[1] To us general heliotherapy does not appear to have any therapeutic value when it is practiced without medical supervision, as is so commonly true at the present time, and it is not devoid of serious risks.

for example, any loss of complete extension of the knee, as revealed by passive hyperextension requires the use of plaster shells.

The use of moulded supporting apparatus and especially of plaster shells rests on knowledge of the mechanisms of defective postures. Under the influence of pain, chronic articular inflammation ends by causing a spasm of the periarticular muscles. And it is usually the flexor groups that overcome the extensors. As soon as an extremity has begun to give way, it can no longer find a position of rest, even when the patient is in the recumbent position. And thus a vicious circle is established which tends to aggravate the defective posture. By keeping the extremity perfectly in the position that it had assumed, the plaster shell relieves the spasm in a few days and breaks the vicious circle. After a few days of complete immobilization, stiffness and ankylosis will be avoided by taking the extremity out of the shell once or several times a day, and by a few active movements which should be made slowly but completely.

Material

The preferable material is a plaster bandage that sets rapidly and permits perfect and regular moulding and produces solid and light casts. This type of bandage, by producing light shells, is especially useful for the upper extremities.

Preparation of the Patient

Several days before the immobilization he should be put at rest and he should take analgesics during the hour preceding the application of the cast.

Postion in which the Plaster Cast should be Applied

For the application of the cast the affected joint should be in the position of function or as near this position as can be obtained without force or trauma.

The shell should be obtained from a circular cast that will immobilize the joint from three to six days. Better results are obtained with complete immobilization for a few days than with an immediately prepared bivalve cast: during this short time stiffness does not increase and muscular tension is much more completely reduced.

The circular cast should be cut into two halves and thus made removable, only the posterior half will be used as a shell, and this will permit daily mobilization of the joint

At first the extremity should be taken out of the shell for a few minutes each day, and active mobilization should be practiced Thus, thanks to this immobilization interrupted by exercises, the patient will obtain, not ankylosis, but freedom of the joints.

Positions in Which the Different Joints Should Be Immobilized and the Different Types of Casts That Should Be Applied

The Spine

In Ankylosing Spondylitis the spinal column has a tendency to bend forward To prevent this all that is necessary is to place the patient on a hard bed To preserve lumbar lordosis a pillow may be put under the back at this level.

Rarely and only in a few very painful cases, we prepare a plaster bed consisting of a large plaster shell This should be moulded with the patient lying on his belly, and it should extend from the sacral region to the shoulders

Swaim Corset with Sternal Support. Wearing this corset is the best way to maintain or to improve the position of the spine, it will be considered with the methods of correcting defective postures (p 327)

Lower Extremities

The Hips Flexion, with adduction of the hips, is the position that hips affected by Ankylosing Spondylitis tend most frequently and spontaneously to take

The functional posture of these joints is extension, an intermediate position between external and internal rotation, and the intermediate position between adduction and abduction In cases with bilateral involvement it is preferable to place the hips in a position of abduction rather than in the intermediate position, that is, with the lower extremities parallel, because the intermediate position would make walking impossible and would embarrass the visceral functions to the maximum degree

The plaster shell consists of a large gutter enclosing the pelvis and the two lower extremities down to and including the feet. This

cast is also employed after the hip has been replaced into a good position under general anesthesia, a procedure which we have had little occasion to use in Ankylosing Spondylitis.

The Knees The chief danger is loss of extension, which interferes with walking or makes it impossible This joint, therefore, should be immobilized in the position of maximum extension

The cast should include the lower extremity from the crotch to and including the foot. Usually gutters that stop above the malleoli are not well tolerated, the lower edge pressing disagreeably on the Tendo Achilles and on the projecting malleoli.

The Ankles and Tarsi. When an ankle is affected by an inflammatory process, it tends to assume a position of plantar flexion with equinism, if care is not taken, the weight of the foot and of the coverings will increase this deformity Laterally the foot tends to deviate either in varus or, more frequently, in valgus When the vertical position is resumed, the patient will find it very difficult to walk.

Therefore, the position of the ankle in the cast should be a slight plantar flexion, at a 95° angle or, at most, at an angle of 90°, the foot should be in a position intermediate between abduction and adduction

The cast should support not only the ankle but the entire foot down to the ends of the toes, and it should extend upward as far as the upper third of the calf

To correct deformities of the toes is difficult, and at the present time we have not yet found a satisfactory solution of this problem.

Upper Extremities

The Shoulders The functional position of the shoulder is in abduction to 45° and in slight external rotation The inflamed shoulder, on the contrary, tends to rest in adduction and internal rotation Almost never do we attempt to apply plaster casts to shoulders affected by Ankylosing Spondylitis we confine our measures to putting a pillow between the thorax and the arm in order to try to preserve abduction

The Elbows The upper extremity serves for prehension (carrying the hand to the mouth and head in eating or combing the hair), and these movements depend on the mobility and position of

the elbow To preserve this function, the elbow must be in a position of flexion at less than 90°, loss of extension, on the contrary, interferes very little with this function Inflammatory diseases tend to fix the elbow between complete extension and a right angle, thus entailing a large measure of disability (the patient must depend upon others to eat, to shave, etc)

In order to obtain a good result the elbow should be placed in flexion at an angle of 85° or 80° and at the same time the forearm and hand are placed in semi pronation, with the palmar surface turned toward the thorax.

The cast extends upward to the upper third of the arm, and downward to the styloid processes of the radius and ulna, these processes serving as points of support to provide a good immobilization Over the outer surface of the arm and the upper surface of the forearm the plaster shell should be open in such a way that, when the elbow is in the splint, it will be prevented from performing movements of flexion and extension

The Wrists The functional positions of the wrists are many, but the position which, to us, seems most useful is one of slight hyperextension, a patient with the wrist stiffened in this position, without lateral deviation, can still use his fingers.

The Metacarpo-Phalangeal and Phalangeal Joints These being rarely involved in Ankylosing Spondylitis, they should be put in slight flexion to facilitate prehension In these cases the cast should not be circular it should consist of a curved shell including the palmar surface of the hand and extending from the ends of the fingers to the upper third of the forearm

Medical Exercise

The fear of "articular ankylosis" leads the patients not to rest in bed and even to 'walk' on inflamed joints.

Nowadays, with a reasonable system of bed exercises, this fear is completely unwarranted when the principles of "rest and movement' are applied, not only is joint mobility preserved but a favorable action on the inflammation of these joints is also obtained, hence the importance of regular exercises for all bedridden patients.

Principal Physiologic Actions

a. Exercises maintain a normal amplitude of movement by rendering the capsule more supple and preventing the formation of adhesions

b They maintain muscular trophism and prevent the retraction of muscles and tendons.

c This is the best means of preventing decalcification of bones, which does not seem to be influenced by injections of calcium and of other so-called recalcifying treatments

d The patient preserves a good general condition from improvement in the blood and lymphatic circulation as well as in pulmonary respiration

General Principles of These Exercises

a The types of exercises that should be employed in Ankylosing Spondylitis are

Exercises of static muscular contraction,

Active assisted exercises

Active free exercises

b They should be practiced two or three times a day from five to 10 minutes, and they should be done on a hard bed

c. A small number of very simple movements (a maximum of 3 for each joint) should be prescribed At first each movement should be repeated five times, and then the number should gradually be increased to 15 to 20 times

d They should be done slowly, to the full extent, but without force

e When at the end of the exercises, there is an increase in pain swelling or stiffness, they should be reduced or suspended for a few days, and they should be resumed two or three days later, but in more moderate fashion

f At the beginning the exercises should be taught and supervised by a physician or a medical assistant

The patient is given definite written instructions concerning the different movements to be executed

Spinal Exercises Postural Exercises

The patient is stretched on the back, without any pillow for the head but with a pillow under the thorax, another pillow under the

knees the hands are crossed back of the head, the elbows tending to touch the plane of the bed This position should be maintained from 20 to 30 minutes after each meal (position 1)

The patient is made to lie on the belly with the legs extended, the arms over the head, and a pillow under the abdomen After this exercise, he should be required to hold this position from 20 to 30 minutes twice a day (position 2)

Respiratory Exercises The patient being in position 1, he should take a full inspiration and, raising his chest, he should expire as completely as possible.

Every week, the patient's respiratory capacity (expansion) should be measured in centimeters.

OTHER EXERCISES FOR THE SPINAL REGION Lying on his belly, the patient should contract the gluteal muscles and maintain this contraction for a few seconds, and then relax

Lying on his belly, with the thorax and head quite flat, the patient should raise the legs with the knees extended

Lying on his belly, with hands on hips, he should raise the head and shoulders from the plane of the bed and keep this position for a few seconds

Exercises for the Hips

The patient should stretch flat on the belly on a hard bed morning and night for 10 to 15 minutes

Every morning the patient, lying flat on his back, should make a few limbering movements of the hips

a Flexion and extension of the thighs on the pelvis.

b With the knee and hip semi flexed and with the heel resting on the bed, the patient should separate the thighs and should bring them together again

c With the lower extremity extended on the bed the patient should rotate the lower extremity and foot inward and outward

Each of these movements should be executed slowly and completely from 10 to 15 times.

Exercises for the Knees

With the patient sitting on the edge of his bed, his feet dangling and his arms crossed over his chest, he should successively raise one foot and then the other while trying to obtain maximum extension

of the knee This puts his quadriceps muscle to work without "crushing" the inflamed articular surfaces.

Indications Between acute attacks and for all bedridden patients these exercises should be practiced and gradually increased

For patients in plaster shells these exercises should be compulsory, they should be started three days after the application of the shell, and they should be continued until the patients resume their activity

Exercises in Water The exercises may be facilitated if the patient is wholly immersed in warm water, this can be achieved especially in pools of warm water

Massage

Massage has a limited role in the treatment of Ankylosing Spondylitis It should be entirely muscular, it should not touch the affected joints and it should be avoided during painful episodes.

Thus massage may be useful to combat muscular atrophy, but it will only serve to complete the action of exercise for which it should never be substituted

Analgesic Agents

They provide good rest for the affected joints and facilitate their 'derusting,' that is, thanks to their use, orthopedic precautions and exercises can be practiced more effectively

Aspirin

Usually we prescribe *Aspirin* in tablets of 0 50 g without any restriction on the quantity to be taken generally the patients take one tablet in the morning, before rising, to facilitate limbering the joints, and one tablet at night for better sleep

When Aspirin is not well tolerated by the stomach the patient should take a derivative of Acetylsalicylic acid or an alkaline mixture which the stomach is more likely to tolerate

Amidopyrine

Patients seem to tolerate this drug well We use it in doses of 0 30 g three or four times a day Since this drug tends to accum-

ulate slightly in the body, it should be given interruptedly during the week for example, five days a week, followed by two days of rest When given in this manner the drug may be administered for months or years We have not observed certain ill-effects in the blood (agranulocytosis) which have been reported from the United States Because of the possibility of an idiosyncrasy to this drug, the physician should first make sure that the patient will tolerate it well In one instance which, fortunately, was an isolated case, a few drops *by mouth* were sufficient to provoke an immediate and severe syncopal condition

Butazolidine and Irgapyrine

These drugs are much used because they render great service during exacerbations of pain They appear to act not only on the pain, but also on the inflammation They may be combined with Gold treatment for, when they are given at the beginning of a course of treatment with Gold salts, they bring about rapid improvement

Irgapyrine (a solution of Phenylbutazone and Pyramidon) is given by way of the digestive tract (capsules or suppositories) or intramuscularly For the last a 20% solution is employed The usual injection of 3 c c is given daily or every second day A series comprises from 10 to 12 injections. Before starting the treatment, the physician must assure himself that the kidneys are functioning normally and that the blood count is normal

Butazolidine (Phenylbutazone) may be employed by the same routes as Irgapyrine It has the advantage over Irgapyrine of not containing Pyramidon

Other Analgesics

In a few cases in which the pain is very severe it is necessary to prescribe more energetic analgesics *Codeine* in doses of 0 02 g in each capsule, with or without *Amidopyrine*, and *Dolosal* subcutaneously (2 c c. in 2 or 3 daily injections) or in the form of suppositories This is well tolerated even when given over long periods.

As in all rheumatic diseases, the use of *Morphine* should be strictly forbidden. When one has observed a few cases of Spondylitis or of Rheumatoid Arthritis in which some physician was weak enough

to prescribe morphine, he is bound to have for it a profound horror The prolonged use of morphine in a disease that does not kill its victims, brings on a state of stupidity and depression of personality which every physician should regard it as a duty to avoid at all costs

BASIC TREATMENT

Since the object of internal treatment is to stabilize the disease, it should be carried out as long as signs of progression continue. But present-day methods of treatment cannot be employed con *tinuously for months or years they should be given in interrupted* series, that is, with a certain rhythm When drugs or other medication are given in this manner, they are well tolerated and may be continued longer than when the treatment is continuous These remarks are as valid for radiotherapy as for medicinal prod ucts.

Since the usual progress of Ankylosing Spondylitis is punctuated by exacerbations of variable duration, it is very difficult to judge the effectiveness of any given medication We will mention here only therapeutic agents that have proved their value, and espe cially those whose effects we have personally followed for many years. Cortisone and A C.T H as they are now employed, have a spectacular action, that is, an immediate and pronounced, but transitory, action Only experience will permit us to determine the most effective dosage and their indications.

RADIOTHERAPY

In 1930 Kahlmeter was the first to observe the effectiveness of roentgen treatment in three patients who were afflicted with Anky losing Spondylitis (114) In 1935 G Scott (190) explained his technic of roentgen treatment with 'large fields' and the good results that he had obtained in cases of Ankylosing Spondylitis and in subsequent communications he drew attention to this method

Since then other writers have published the results obtained with roentgen treatment. In 1941 Smith, Freyberg Peck and Lampe (204 205) adopted this method and gave details about the results

obtained, after having carried out clinical and biologic examinations before and after treatment

At the present time roentgen treatment is considered as one of the most effective methods of treatment for Ankylosing Spondylitis

Technic

The technics vary greatly, each therapeutic radiologist applies his own method, and this diversity of technics has had something to do with the rather limited use of this method Nevertheless, on the whole, three types of roentgen treatment may be distinguished

Semi-Penetrating Roentgen Treatment with Large Fields (Scott's Method) (192)

PENETRATION 100 to 120 KV, 4 milliamperes.

FILTER 3 mm of Aluminium

DISTANCE 50 cm.

FIELD At one session of a course of treatment the upper half of the trunk is irradiated, then the lower half

DOSE. 60 roentgens are given to each of these two fields (only 30 roentgens are given at the first sessions)

RHYTHM OF THE SESSIONS OF TREATMENT Once a week for six weeks, with a total dose of 660 roentgens at each course of treatment

The course of treatment may be repeated after three months' rest.

Semi-Penetrating Roentgen Treatment directed toward Localized Fields over the Painful Segments

We give here the technic now used by Freyberg and which, to us, seems one of the best planned methods (91)

PENETRATION 140 KV, 10 milliamperes

FILTER 0 5 mm of Copper and 1 0 mm of Aluminium.

DISTANCE 50 cm

FIELDS *Sacroiliac field* 12 × 18 cm.
Lumbar field 18 × 16 cm
Lumbar field 18 × 16 cm
Cervical field 14 × 12 cm.

Additional fields are treated in cases with involvement of the hip or shoulder

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Since then other writers have published the results obtained with roentgen treatment. In 1941 Smith, Freyberg, Peck and Lampe (204 205) adopted this method and gave details about the results

obtained, after having carried out clinical and biologic examinations before and after treatment

At the present time roentgen treatment is considered as one of the most effective methods of treatment for Ankylosing Spondylitis

Technic

The technics vary greatly, each therapeutic radiologist applies his own method, and this diversity of technics has had something to do with the rather limited use of this method Nevertheless, on the whole, three types of roentgen treatment may be distinguished

Semi Penetrating Roentgen Treatment with Large Fields (Scott's Method) (192)

PENETRATION 100 to 120 KV, 4 milliamperes

FILTER 3 mm of Aluminium.

DISTANCE 50 cm

FIELD At one session of a course of treatment the upper half of the trunk is irradiated, then the lower half

DOSE 60 roentgens are given to each of these two fields (only 30 roentgens are given at the first sessions)

RHYTHM OF THE SESSIONS OF TREATMENT Once a week for six weeks, with a total dose of 660 roentgens at each course of treatment

The course of treatment may be repeated after three months' rest.

Semi-Penetrating Roentgen Treatment directed toward Localized Fields over the Painful Segments

We give here the technic now used by Freyberg and which, to us, seems one of the best planned methods (91)

PENETRATION 140 KV 10 milliamperes

FILTER 0 5 mm of Copper and 1 0 mm. of Aluminium

DISTANCE 50 cm

FIELDS *Sacroiliac field* 12 × 18 cm.
Lumbar field 18 × 16 cm
Lumbar field 18 × 16 cm
Cervical field 14 × 12 cm.

Additional fields are treated in cases with involvement of the hip or shoulder

THE DOSE PER FIELD (measured in air) 150 roentgens at each session, each field is irradiated three times and thus receives during each course of treatment 450 roentgens.

RHYTHM OF THE SESSIONS The sessions are repeated every two days, and at each session two fields are exposed

At the first, third and fifth sessions, fields 1 and 3

At the second fourth and sixth sessions, fields 2 and 4

Each course of treatment requires two weeks

REPETITION OF THE COURSES A second course is given after five weeks rest, with smaller doses 100 × 3 = 300 roentgens to each field In the event that pain should recur later, more sessions of treatment, with a dose of 100 roentgens to each field, are given.

It should be mentioned that this technic is but a modification of the technic previously employed by Smith Freyberg and Lampe (205), which, because of excessive doses, was sometimes not well tolerated In their new technic we find a tendency to use 1) a lower voltage (formerly 220 KV, and now 140 KV), 2) larger fields so as to irradiate the paravertebral muscles 3) smaller doses (formerly 600 roentgens, and now 450 roentgens at each course)

Roentgen Treatment of the Pelvis *This is employed only in* men by J Forestier and P Robert. To these authors it seemed indicated that the true pelvis, which is contiguous to the sacroiliac joints (the first spinal joints to become involved), should be irradiated The roentgen tube is aimed in such a way that the rays "lose themselves" in the true pelvis (base of the bladder, seminal vesicles, prostate gland)

PENETRATION 160 to 180 KV, according to the obesity of the patient

FILTRATION 0 5 mm to 1 0 mm. of Copper

DOSE 100 to 180 roentgens at each session of treatment. Sessions repeated every two or three days

NUMBER OF SESSIONS Eight, the total dose in each course of treatment is thus from 1500 to 1600 roentgens.

In cases in which the spinal manifestations are pronounced, irradiation of the pelvis may be combined with irradition of the spinal column with a technic approximating that which has just been described

Tolerance

For the patient deep roentgen therapy may be accompanied by disturbing or even dangerous secondary effects.

According to Smith, Freyberg and Lampe (205), the most important of these effects is *leucopenia* This usually supervenes after too large doses or during a second course of treatment A slight and transient reduction in the number of leucocytes is common Marked leucopenia is rare, of 52 treated patients only three have had very low leucocyte counts 3,400, 1,700, and 1,000 white cells In no case did the leucopenia have any serious consequences, and the number of white cells returned to normal within a month No case of leucopenia below 3,000 leucocytes was noted with Freyberg's technic of small doses (91), which we have described

When the lumbodorsal region was irradiated, *digestive disturbances* consisting of anorexia, nausea and vomiting were observed Usually these symptoms begin from four to six hours after a session of treatment, and they completely disappear in two or three days. With the moderate doses now used they have occurred much less frequently than heretofore.

Cutaneous accidents have not developed Pigmentation of the skin in the treated fields is observed and, according to P Robert this is more marked than in patients who are not afflicted with spondylitis A temporary loss of hair may be noted when the cervical region is irradiated It is wise, therefore, to irradiate below the scalp line

In women large doses applied over the hips may result in an arrest of menstruation (the ovaries are situated near the bottom of the acetabulum) and may be followed by sterilization Irradiation of the sacroiliac joints is less dangerous In women, by all means, a total dose of 300 roentgens over the sacroiliac joints should not be exceeded it is even wiser to limit one's self to smaller doses In men secondary reactions in the genital organs do not occur

Incidents or accidents, therefore, arise from too large doses at each session, from too long a course of treatment, or from courses of treatment repeated too often As far as we are concerned, we have observed only slight incidents, and this only in a small proportion of cases.

Action of Roentgen Treatment

According to the authors cited, the *immediate results* were good in 70 5% of their 52 cases (205) The results were especially favorable in the cases in which the roentgenographic changes were confined to the sacroiliac joints (92% of good results)

Pain is the symptom which is most definitely influenced by roentgen treatment A distinct abatement of pain occurs rapidly, and this abatement may be observed within a few hours after the first session but most frequently after the first week of treatment In other cases the abatement is less distinct and is noted only after the second course of treatment.

Improvement in spinal mobility (finger to-floor distance and thoracic expansion) depends on the phase of the disease in which roentgen treatment is given This improvement may be marked in cases in which the stiffness is due only to pain and to muscular spasm then the patient recovers normal mobility as soon as the pain has disappeared

In the hips, involvement of which is often accompanied by severe pain, roentgen treatment may be very effective As for the other peripheral joints, roentgen treatment seems to have a variable effect.

The rate of sedimentation was favorably influenced in 41% of the cases (205) In a few cases it returned to normal the general condition also improved

Long Range Results Personally we do not have any cases that have been treated exclusively with roentgen rays and that have been followed for many years

One series of patients who were observed for a very long time and who were treated only with deep roentgen therapy is that of Robinson (170)

Before the first course of treatment, 55 patients, who had been followed for four five, and nine years, were divided into three groups according to the extent of the roentgenographic changes

Group 1 18 patients presented roentgenographic changes that were confined to the sacroiliac joints Group 2 24 patients had lumbar or dorsal involvement Group 3 13 patients had general involvement of the spine

Group 1 out of 18 patients eight obtained a complete remission, some for eight years, others for five years Seven patients had recurrences and were again treated successfully Objective improvement in stiffness was obtained in 13 patients Nevertheless, serial roentgenograms indicated definite progression of the disease in more than half of the cases

Group 2 out of 24 patients, 11 obtained a complete subjective remission and 7 others obtained incomplete relief from pain Mobility of the spine improved in 15 patients definite progression of the roentgenographic changes was found in 21 of 24 patients

Group 3 out of 13 patients, 11 obtained a certain degree of relief from their symptoms improvement in mobility was noted in only three patients An increase in the roentgenographic changes was found in all cases. The variation curve of the sedimentation rate was not recorded

This detailed study demonstrates that, in spite of the remission of the pain syndrome that may rapidly be obtained with this method of treatment, the disease continues to progress anatomically, and this is proved by serial roentgenograms of these patients

This would seem to prove that roentgen therapy has an especially analgesic action The rapid suppression of pain permits these patients to perform gymnastic exercises which tend to preserve the mobility of joints, and even to carry on some social activity

Indications

This kind of treatment is indicated in all cases in which pain in the spine is a prominent feature

In cases with local pain in the spine, treatment through a single field may be sufficient This field should not be confined to the painful region but should also include the adjacent segments In cases with diffuse pain in the spine roentgen treatment should be directed through 3 or 4 fields

Thorium X[1]

Thorium X, which was first used by Léri, is a radioactive agent

[1] A general review of radioactive medication in the treatment of rheumatic conditions and of its dangers was published in 1937 by F Coste (48). This review includes an abundant bibliography From this communication we have taken a few excerpts.

with an atomic weight of 232 12 from it can be obtained an active substance called radiothorium which has a half life of 1 90 years. At one time the direct therapeutic use of radiothorium "radithor" was rather widespread in America, but later its use was abandoned because of numerous accidents

Thorium X is obtained from a hydrobromic or hydrochloric solution of radiothorium which is precipitated, and then filtered This radioactive solution emits only α rays, that is, atoms of helium with a high ionizing power but with little penetrating power and with a short life The activity of Thorium X decreases rapidly its half life period is 3 64 days it becomes transformed into a series of compounds with a very short life (thoron, etc) When the Thorium X is pure and free from any trace of mesothorium, a condition which present-day methods of preparation attempt to achieve, its activity drops to zero after three or four weeks The short life period of Thorium X and of its derivatives thus renders impossible any accumulation in the body and enables one to use it like a drug

Nevertheless, the rate of disintegration also constitutes a danger, because the product liberates its energy in an almost explosive manner According to Lacassagne (132) the short contact period of Thorium X with the tissues does not eliminate the possibility of late accidents, and even of immediate accidents

The radioactive solution which does not contain radiothorium, is mixed with a solution of sodium chloride in order to make it isotonic and thus injectable

The activity of this solution is measured by comparing the α radiation with that of a definite quantity of radium taken as a standard and expressed in micrograms Knowing the half life period we know the dose that will be contained by the ampoule on a given date which is inscribed on the ampoule and on which the injection is to be made The commercial preparation of the product is made on a set day each week.

W Koch has shown experimentally that Thorium X is electively deposited in tissues undergoing calcification or ossification This agent has the property of retarding or arresting the development of

the ossifying process, either in young persons or in cases of pathologic ossification, hence its therapeutic usefulness

Mode of Employment

In an isotonic aqueous solution, it should be injected intravenously or, if this cannot be used, intramuscularly In order to eliminate any danger of local reactions, the injections should be deep, and the points of injection should be changed

In two patients who had received injections of Thorium X without these precautions having been observed, we have noted a circular dermatitis in the buttock, where the injections had probably been made too superficially

Another patient who, when Thorium X first came into use, had given to himself, without a prescription, about twenty subcutaneous injections of large doses into the outer surface of the thigh and always at the same spot, developed a radio-dermatitis with ulceration, which has persisted for 15 years with only temporary remissions.

Dosage

We give a weekly injection with gradually increasing doses

50 μg twice

100 μg twice

150 μg twice

200 μg from two to four times, making the total dose for the series 1,000 to 1,300 μg

Shorter series of 1,000 μg or longer series of from 1,500 to 2,000 μg may be given according to the severity of the disease Thus, in one case with few painful symptoms, a series of from 1,000 to 1,200 μg may be given In severe cases with marked symptoms stronger total doses of 1,200 to 1,500 μg may be employed Courses of treatment with a total dose of 2,000 μg should be given only in cases that have proved very resistant to other forms of treatment and that, although the patients' general condition is good, have shown, in previous courses, satisfactory tolerance of the product

After each course of injections, a rest period of four months should be allowed, that is, the patient may receive, at most, two series of injections a year

Tolerance

It is admitted that no trace of Thorium X is found in the blood 24 hours after it has been injected It is deposited in large measure in the bones and in smaller measure in the spleen kidneys, and liver It is eliminated in part through the kidneys and intestine Since the product and its derivatives have a short life, their activity, after three or four weeks is nil

Theoretically danger from its use does not arise so much from its accumulation as from the violence of its liberation its energy may produce cellular changes, especially in young and poorly differentiated cells such as the mother cells of the hematopoietic organs and those of the genital glands

Theoretically accidents may be of four kinds

Blood Accidents In 1937 Coste (48) found in medical literature five cases of severe blood accidents from a total number of patients estimated in France at more than 15,000 These accidents had occurred in cases in which the patients had received from four to seven injections of 500 to 650 µg each or in two other cases, fifty five injections of 300 µg, that is, enormous doses Out of 91 of his patients (48) who had received from one to five courses of 700 to 800 µg Coste had observed in seven cases general fatigue and a reduction in the number of leucocytes or erythrocytes, which had forced him to stop the treatment.

In our patients we have observed this tendency to leucopenia and anemia, accompanied by general fatigue, but these disturbances have never been marked, and during the past 20 years we have not encountered a single grave or marked accident as far as the blood has been concerned

Local Radiodermatitis This form of dermatitis, which occurs at the point of injection of the radioactive agent, develops only in cases in which intramuscular injections have been given A few cases have been published in the literature (132) Two examples of our own have already been cited they were due to faults in technic that could easily have been avoided

Bone Necrosis Four cases have been reported in the French literature These were cases of osteonecrosis of the mandible in patients who had previously been treated with Thorium X Such accidents are very rare and have been caused by much larger

doses than those that are usually employed In two of the reported cases Thorium X could not be found in the necrosed bone

Osteosarcoma There has not been a single case of osteosarcoma that could be attributed to Thorium X (48)

Since 1937, no cases of accidents have been reported

To sum up, Thorium X, in moderate doses, may produce a slight leucopenia, a reduction in the number of erythrocytes, and slight general fatigue Radiodermatitis is a possible complication in cases in which Thorium X is injected intramuscularly with a bad technic Bone necrosis due to Thorium X has not been proved We believe these rare late accidents may be due, not to Thorium X itself, but to the presence of Mesothorium, which is a product with a long life The case observed by Mme Laborde (128) is very striking, and it seems very difficult to obtain pure preparations of Thorium X

Personally, we have administered this substance, during the past 20 years, to more than 200 patients (usually in several courses), and we have never had a single accident on the contrary, we have noted a few transient incidents

1) A temporary return of pain may be observed after each injection, this return of pain has never been marked and has not interrupted the treatment On several occasions (6 or 7 times) we have noted an exacerbation of iritis after the first injection of Thorium X in patients who had already had iritis.

2) General fatigue sometimes rather marked has been noted after a course of treatment, and this has continued for a few months. In spite of this fatigue, since blood counts showed only a slight leucopenia the treatment of these patients has been continued with good results

Results

We have treated more than 200 patients with one or more courses of Thorium X

Out of 41 cases in which the patients were treated exclusively with Thorium X and in which they were followed from two to 10 years, we have noted

Improvement in 32 cases transient improvement (from 6 to 9

months) in 10 of these cases, and persistent improvement in the others

No improvement in nine cases.

Thorium X has brought about improvement in three fourths of the cases which we have treated But it must be admitted that, in these cases, the intensity of the disease varied and, as was shown on page 186, it is possible that the improvement of some of them may have coincided with a spontaneous remission, the duration of the attacks varied considerably, and the treatment was spread over many months.

The improvement manifested itself by

1) Pain in the *spine and radiating pain diminished or disappeared* Often, after an improvement of from six to nine months following the first course of treatment, a fresh attack of pain supervened, this indicates that the second course of treatment should be given soon enough after the first course to prevent these recurrences

2) Stiffness of the spine when this was related to muscular contracture which, in turn, was related to the pain, the stiffness improved but most frequently the stiffness continued or increased, even in cases in which the pain had diminished or in which the general condition had improved

3) The involvement of peripheral joints is only slightly influenced by treatment with Thorium X.

4) The patients' general condition has often been favorably influenced, with recovery of the lost weight and appetite. This improvement seems to be related to a direct action of the medication on the disease process and is not a simple consequence of relief from pain

5) The sedimentation rate has improved, in some cases, in proportion to the clinical symptoms, but in other cases this rate has not improved in the same way

Indications

Thorium X is indicated in all cases of Ankylosing Spondylitis, and especially in those with pain in the spine or with radiating pain It has also given us good results in cases of Ankylosing Spondylitis with psoriasis.

It is contra indicated only in cases in which the general condition is very bad and the blood count abnormal, and in those in which the patients' antecedents show some disease that could have weakened the hematopoietic system

Radon

Radon is an inert gas that results from the disintegration of radium It is in the form of a solution that radium salts most readily liberate radon

Its half life period is 3 25 days, it is therefore similar to that of Thorium X, which is 3 64 days Radon gives off 92% of α rays, and 8% of β and γ rays Therefore, it differs from Thorium X which emits only α rays.

Vaugeois, under d'Arsonval's direction (2), found the most practical way of obtaining radium emanation, or radon, to make it injectable in a gaseous atmosphere and to utilize it in medicine Instead of a solution of radium, which is difficult to handle outside of a laboratory, a solid emanation source of small volume has been utilized This emanation source, which is composed of a radium salt encased in a porous colloidal mass, is enclosed in a metallic tube This tube emits, in unit time, the quantity of emanation for which it is designed

At each end of the tube is fixed a valve the gas in the tube becomes charged with radioactive emanation while the valve is closed A table enables one to know the quantity of emanation that has accumulated in the tube during a given period that the tube has been closed To one of these valves is fitted a syringe with a capacity greater than that of the emanation tube (in practice, 20 cu mm.), and this is charged with air, at the other end of the tube is fitted a needle for subcutaneous injections after the needle has been inserted and after the valves have been opened, the gas is slowly injected

Sometimes the nitrogen of the air is not well absorbed by the tissues and a prolonged subcutaneous emphysema results In this event the atmospheric air should be replaced by oxygen or by carbon dioxide

Dosage

Usually we use tubes which, after being charged for 24 hours, contain 10 microcuries of radon

A subcutaneous injection is given every two days, with a total of from 10 to 12 injections at each course of treatment, or from 200 to 240 microcuries at each course In general a maximum of two courses of injections are given each year

Tolerance

Radon undergoes transformation into a succession of products *with a very short life (a few minutes) radium A, radium B, radium* C the first products with a long life which it emits are radium D and Polonium Radon does not combine with any other substance and thus does not yield any insoluble compounds.

Four hours after an injection, traces of it are no longer found in the blood It is eliminated through the respiratory tract. In practice we have never observed any accumulation of radon or of *its derivatives with a long life The theoretical possibility has been* calculated, and it is insignificant Its toxicity might arise from its rapid destruction but this is tempered by its rapid elimination Radon in the doses which are usually injected is completely harmless

We have not observed any signs of intolerance.

Results

We have used radon in about 100 cases. Since most of our patients have received, at different times, either radon or some other radioactive treatment, it is impossible for us to report definite long term results

We have noted a definite influence on pain and on the general condition In these respects improvement may be transitory (a few months) or persistent We have not been able to determine its influence on the sedimentation rate It has not seemed to have any action on peripheral involvement

Indications

The indications for radon seem to be the forms of the disease in which pain in the spine is an outstanding factor, in which the pain is refractory to other kinds of treatment and in which the rate of

sedimentation is normal or nearly so For reasons of convenience we have alternated the use of radon with that of Thorium X

Gold Salts

The use of gold salts which was introduced by J Forestier for the treatment of rheumatoid arthritis, has been tried by him in other forms of inflammatory rheumatism and especially in Ankylosing Spondylitis, originally it was employed in cases with marked peripheral manifestations, and later in cases with spinal involvement alone

Usually we employ *organic* salts containing from 30% to 50% of the metal either water soluble 1) thiopropanol sulfonate of gold and sodium (allochrysine), 2) auro-thio-malate of sodium (Myochrysine), or insoluble salts to be used in an oily suspension (oleochrysine, myoral), *more rarely mineral salts* that are soluble in water thiosulfate of gold and sodium (sanochrysine)

We prefer the water soluble organic salts, because gold salts in an oily suspension may cause abscesses, because of the ease of injection, we prefer the intramuscular route We reserve the mineral salts, (sanochrysine), which must be injected intravenously, for cases in which the organic gold salts do not seem to have any effect.

Dosage

The doses are the same as those used in the treatment of rheumatoid arthritis

Dose at each injection 0 10 g (0 05 g should be given at the two first injections of the first course of treatment)

One injection each week,

A total of 10 or 12 injections, that is, 1 0 g or 1 20 g of gold salts at each course Practically speaking we do not modify this dose Patients who have received several courses of this treatment and who have tolerated it well but in whom the gold salts seem to be less active than previously, receive weekly doses of 0 20 g, a course of treatment consisting of only six injections, in such a way that the total dose per course remains the same, but is given in a shorter time.

The interval between the first and second courses of treatment is six weeks the intervals between the subsequent courses of treatment are two months, as long as signs of progression continue Later, maintenance courses will be interrupted by an interval of three or four months.

Tolerance

In nearly a third of our cases of Ankylosing Spondylitis, in which the patients have been treated with gold salts, these have not been well tolerated, but since, in eight cases out of 13, the incidents were transitory, the gold treatment was resumed later

Cutaneous and mucous incidents itching, eruptions, eczema, aphthae, gingivitis Save in two cases of severe Ankylosing Spondylitis, the eczemas have been local and of short duration,

Digestive incidents colic, diarrhea,

Hepatic incidents,

Renal incidents transient albuminuria,

General fatigue (impression of exhaustion)

Blood accidents (agranulocytosis, purpura) are possible, but we have never observed them in our patients who were afflicted with Ankylosing Spondylitis.

Precautions to be Followed

Before starting gold treatment, the physician should

1) *Look into the patient's pathologic antecedents from the* point of view of the blood the kidneys, and the liver When these have been present, laboratory examinations should be made blood count, platelet count, bleeding time and coagulation time.

2) Systematically examine the urine for albumin

3) In patients who have had eczema or urticaria, before a course of gold salt injections is started, from 5 to 10 ampoules of *from 2 to 5 c.c.* of total hepatic extract should be injected preventively, and besides, on the eve of each injection of gold salt, an intravenous or intramuscular injection of sodium thiosulfate should be given

During the Treatment

In the event that an alarm symptom develops, further injections should not be given, their repetition under these circumstances may be responsible for more serious accidents

Most frequently the simple fact of stopping the medication causes a more or less rapid disappearance of the symptoms of intolerance to gold In severe cases two drugs are now available 1) sodium thiomalate intramuscularly 1 ampoule of 2 c c. of a 50% solution is injected once or twice a day during the next few days, then the injections are given at longer intervals when the symptoms begin to recede When the product is given intravenously, it is diluted in five times its volume of water distilled, and injected slowly, 2) the British antilewisite (B.A L) This product, the 2-3 di mercapto-propanol, has the property of combining in the body with metallic gold and of forming a stable complex which is not toxic, and thus of rapidly stopping an intoxication The dose is calculated according to the patient's weight 2.5 mm of B.A L (0 025 c.c. of solution) to the kilogram. An injection is given every four hours, that is, from four to six injections a day during the next three or four days, then two injections a day during the next three or four days, and then a single injection daily for four or five days. The complete treatment should continue about 10 days Symptoms of intolerance may appear malaise, vomiting, and especially local symptoms pain at the point of injection, painful nodules, aseptic abscesses, sometimes with secondary infection These complications are such that this method of treatment should be reserved for serious accidents. But in these cases it should be employed early

Results of Gold Treatment

We have treated 36 cases exclusively with gold salts, and they have been followed for at least two years

We have observed

Transient improvement (a few months) or persistent improvement at subsequent examinations in 15 cases,

A stationary condition in 16 cases,

Aggravation of the disease in five cases

This series is only roughly comparable with that of patients treated with Thorium X, because the gold salts were given especially to patients who presented Ankylosing Spondylitis with peripheral involvement.

The improvement which was noted in a third of the treated cases consisted of a reduction or disappearance of swelling and pain in the peripheral joints, more rarely of a reduction or disappearance of pain in the spine, and of improvement in general condition On the whole, the results are less brilliant than in rheumatoid arthritis but they are not negligible, as have claimed American writers who, doubtless, had not used this kind of treatment with a sufficient number of courses and with sufficiently strong doses.

Indications

Experience has taught us that gold salts should be employed in Cases with involvement of peripheral joints

Cases with a distinct increase in sedimentation rate

Cases in which the disease is resistant to radioactive agents or to roentgen treatment.

Copper Salts

We have employed two organic salts the copper content of which is different

1) Sodium cupro-allyl thiouree benzoate (Cuprilene) (76)

This salt contains 19% of copper, it is used only intravenously in doses of 0.25 g the rhythm of the injections is two or three a week, the quantity of copper salt given at each course of treatment is from 3 to 4 g, that is, from 12 to 16 injections.

2) Methylamine cupro-oxyquinoline sulfonate (Dicuprene) (77)

This salt contains 6 5% of copper and is slightly less active than Cupralene, but it can be given intramuscularly generally the in jections do not cause pain but it is preferable to add to this product 2 c c of 2% Novocain

The injections (0 50 g in each ampule) are given at the rate of three a week for six weeks (18 0 g at each course of treatment)

The interval between the two first courses of copper salt is from four to six weeks, *and from two to three months* between the subsequent courses, that is, intervals similar to those allowed between the courses of gold salts.

The tests of copper salts which we have made in Ankylosing

Spondylitis have not been very encouraging most frequently they have appeared to be inactive In a few cases, nevertheless, we have observed some improvement, in Ankylosing Spondylitis with psoriasis fine results which, unfortunately, were inconstant, were obtained

Copper salts seem to be *indicated* in cases of Ankylosing Spondylitis with psoriasis and in other cases with involvement of peripheral joints, but only after other basic methods of treatment have failed

Cortisone and ACTH

Cortisone (102, 103, 227)

In 1948 the first administrations of Compound E (11-dehydro-17-hydroxycorticosterone) in enormous doses were given intramuscularly by Hench to patients afflicted with rheumatoid arthritis or with Ankylosing Spondylitis

Our own experience is based on about 10 cases which have been observed over a period ranging from six to eight months Doses of Cortisone averaging from 100 to 150 mg daily were given over periods of 20 to 40 days Our results were the same as those obtained by the American authors.

The improvement in the spinal symptoms of Ankylosing Spondylitis is less spectacular than the improvement obtained in the joints of the extremities, which are affected either by rheumatoid arthritis or by Ankylosing Spondylitis, perhaps because these joints are more readily accessible for examination

To us, however, it has seemed that, in cases with effusion of the knees, treatment with Cortisone did not have a favorable effect while the other symptoms of the disease improved Exacerbations of iritis may supervene even during treatment with Cortisone by the general route

After the treatment has been stopped, the symptoms usually have returned This is observed after an interval varying between a few days and two or three weeks Different writers have tried prolonged, continuous treatment with maintenance doses varying from 50 to 100 mg daily (the oral route is best for this method of treatment) But we do not know what may prove to be the remote secondary effects of this hormonal treatment with large doses

To us Cortisone seems to be indicated in very severe cases and especially in cases that present persistent peripheral involvement. Moreover, by relieving pain treatment with Cortisone has facilitated the orthopedic straightening of the spine at the present time, however, we lean toward Butazolidine instead of Cortisone. This form of treatment is contra indicated before bloody orthopedic operations, because it reduces the patient's resistance to surgical shock but is indicated after operation because, by attenuating the inflammatory symptoms, reeducation is facilitated

Cortisone applied locally, or more recently Hydrocortisone (Compound F) (243) does not appear distinctly to have improved the results of arthroplasties which, as is well known often end in fresh ankyloses

Besides the employment of Cortisone during the post-operative period, it may be indicated in cases in which inflammatory symptoms in one or two joints persist while, in order to reduce or abolish the severity of these local symptoms, large doses of Cortisone by the general route would have to be used

ACTH

The adrenocorticotropic hormone or hypophyseal corticostimulin is given in daily doses of 40 to 60 mg , because of its rapid elimination, four injections are given each day The joint effects are similar to those of Cortisone and the therapeutic problems are the same.

Summing Up The introduction of Cortisone and of A C.T H for the treatment of a certain number of diseases, the hormonal origin of which is not apparent, constitutes an important step in medical progress

The diseases which are strongly, but temporarily, influenced are not cured by these drugs which act on the pathologic mechanism of the symptoms and not on their causes These hormones, unfortunately must be employed in considerable doses in comparison with their physiologic level in the body and, when they are given continuously they may be responsible for grave consequences.

In the present state of medical treatment these preparations are interesting means of experimentation and not practical methods of treatment.

Methods of Treatment Advocated As Basic Treatment But, According to Our Experience, of Doubtful Efficacy or None

Shock Treatment (Fever Therapy)

This consists of a series of five or six shocks induced by injecting intravenously gradually increasing doses of a heterogeneous vaccine or peptone The vaccines most often used for this purpose are antityphoid vaccine, antigonococcic vaccine, or a polyvalent vaccine (propidon) The doses should be sufficient to cause a bout of fever between 39 and 40° C. The injections should be given at intervals of three or four days, so that the temperature will have returned to normal before the next shock

We have made little use of this method, its value is open for discussion and, moreover, it causes the patients great fatigue, it also forces the physician to wait two months before giving one of the other basic treatments for example, gold salts, when given immediately after shock treatment, have caused hepatic accidents, such as icterus, more frequently than usual

Tuberculin Treatment

Whatever may be the technic employed (cutaneous, intradermal, or subcutaneous), tuberculin treatment may cause slight focal reactions, without any obvious therapeutic interest

Bogomoletz's Serum, or Antireticular Cytotoxic Serum

Small doses of this serum are claimed to cause stimulation of the reticulo-endothelial system, and the doses used in France are 0 10 0 20 and 0 50 c c intradermally As far as rheumatology is concerned, it has been used chiefly for rheumatoid arthritis, the few cases of Ankylosing Spondylitis that have been treated with this serum have not given encouraging results.

Vitamin D, in Large Doses

This has been used in doses of 600,000 units two or three times a week. This method of treatment has fallen into discredit because of the cases of intoxication that have been published, and also because of the absence of demonstrable results

To us Cortisone seems to be indicated in very severe cases and especially in cases that present persistent peripheral involvement. Moreover, by relieving pain treatment with Cortisone has facilitated the orthopedic straightening of the spine at the present time, however, we lean toward Butazolidine instead of Cortisone. This form of treatment is contra indicated before bloody orthopedic operations, because it reduces the patient's resistance to surgical shock, but is indicated after operation because, by attenuating the inflammatory symptoms, reeducation is facilitated

Cortisone applied locally or more recently Hydrocortisone (Compound F) (243) does not appear distinctly to have improved the results of arthroplasties which, as is well known, often end in fresh ankyloses.

Besides the employment of Cortisone during the post-operative period, it may be indicated in cases in which inflammatory symptoms in one or two joints persist while, in order to reduce or abolish the severity of these local symptoms, large doses of Cortisone by the general route would have to be used

ACTH

The adrenocorticotropic hormone or hypophyseal corticostimulin is given in daily doses of 40 to 60 mg , because of its rapid elimination, four injections are given each day The joint effects are similar to those of Cortisone and the therapeutic problems are the same

Summing Up The introduction of Cortisone and of A.C.T H for the treatment of a certain number of diseases, the hormonal origin of which is not apparent, constitutes an important step in medical progress

The diseases which are strongly, but temporarily, influenced are not cured by these drugs which act on the pathologic mechanism of the symptoms and not on their causes. These hormones, unfortunately must be employed in considerable doses in comparison with their physiologic level in the body and when they are given continuously, they may be responsible for grave consequences.

In the present state of medical treatment these preparations are interesting means of experimentation and not practical methods of treatment.

Methods of Treatment Advocated As Basic Treatment But, According to Our Experience, of Doubtful Efficacy or None

Shock Treatment (Fever Therapy)

This consists of a series of five or six shocks induced by injecting intravenously gradually increasing doses of a heterogeneous vaccine or peptone The vaccines most often used for this purpose are antityphoid vaccine, antigonococcic vaccine, or a polyvalent vaccine (propidon) The doses should be sufficient to cause a bout of fever between 39 and 40° C. The injections should be given at intervals of three or four days, so that the temperature will have returned to normal before the next shock

We have made little use of this method, its value is open for discussion and, moreover, it causes the patients great fatigue, it also forces the physician to wait two months before giving one of the other basic treatments for example, gold salts, when given immediately after shock treatment, have caused hepatic accidents, such as icterus, more frequently than usual

Tuberculin Treatment

Whatever may be the technic employed (cutaneous, intradermal, or subcutaneous) tuberculin treatment may cause slight focal reactions, without any obvious therapeutic interest

Bogomoletz's Serum, or Antireticular Cytotoxic Serum

Small doses of this serum are claimed to cause stimulation of the reticulo-endothelial system, and the doses used in France are 0 10 0.20 and 0 50 c.c intradermally As far as rheumatology is concerned it has been used chiefly for rheumatoid arthritis, the few cases of Ankylosing Spondylitis that have been treated with this serum have not given encouraging results.

Vitamin D, in Large Doses

This has been used in doses of 600,000 units two or three times a week. This method of treatment has fallen into discredit because of the cases of intoxication that have been published, and also because of the absence of demonstrable results.

Sulphur Preparations

In medical practice the use of Sulphur for the treatment of articular diseases is very widespread, and we often have the experience of seeing patients afflicted with Ankylosing Spondylitis who have been treated for a long time with preparations of sulphur

Now it can be stated that these preparations do not have any value for the treatment of Ankylosing Spondylitis and of other forms of inflammatory rheumatism It is useless to prescribe them to patients

Penicillin and Sulfonamides

Some, who believed in the infectious origin of Ankylosing Spondylitis, have used first Sulfonamide or some of its derivatives, and later Penicillin, Streptomycin, or Aureomycin The results have not been striking and we do not feel justified in following this method of treatment.

Synthetic Oestrogens

When these substances have been employed in cases of Ankylosing Spondylitis in men, their efficacy has been very inconstant. Moreover, this method of treatment is accompanied by a few troublesome symptoms, such as a sensation of tension in the breasts with increase in their volume and impotence

Sodium Salicylate

This drug has been greatly used in all forms of rheumatism that are accompanied by articular swelling In Ankylosing Spondylitis it has only an analgesic effect, like Aspirin, with the disadvantage that, when it is given intravenously, it causes sclerosis of the veins and thus prevents the use of other kinds of medication that can be given only through the veins

Treatment of the General Condition

The general condition of the patient plays an important part in the evolution of the disease it is therefore necessary that 1) the patient should be placed in the best possible living conditions 2) that infectious foci (dental abscesses, pulmonary diseases, genital

infections) should be sought for and eliminated, 3) that, when the patient presents a condition of general deficiency, he should receive treatment to increase his ability to take basic treatment

Calcium Gluconate

Given intravenously in a 10% solution, 10 c.c ,

10 to 20 injections in one course of treatment, at the rate of three or four injections a week,

One or two courses of this treatment should be given between each course of basic treatment,

Calcium gluconate may be combined with Vitamin C, of which 2 c c may be injected in the same syringe as the gluconate, or it may be combined with Vitamin D in doses of 600 000 units given orally every eight days as long as the course of calcium gluconate continues.

With this treatment a certain degree of improvement in general condition (weight, sleep, appetite) is observed

Blood Transfusion

In severe forms of the disease we give small transfusions of 150 c.c , which are repeated twice a week, in order to improve the patient's general condition

A recent study (198), bearing on 60 cases of rheumatoid arthritis shows that blood transfusion has a transient effect on the rate of sedimentation, on the hemoglobin content, and on the cellular volume Twenty-eight days after transfusion, the patient's condition returns to what it had been prior to all these laboratory examinations Six months afterwards, the patients who have been treated with blood transfusions may be distinguished from another series of patients who have received the same treatment without blood transfusions by an appreciable improvement in general condition

SPA TREATMENT

At all times so-called rheumatic diseases have been one of the major indications for hydromineral therapy or crenotherapy It was natural that patients who were affected by painful diseases of the spine should seek, at hot springs, relief from their miseries

The hot springs that are useful in the treatment of Ankylosing Spondylitis have a temperature higher than 35° C, and the temperature often rises to 50°, or even higher, usually these springs contain simple water or water with a small percentage of chlorine or sulphur

Among the hot springs in France we will mention Aix les-Bains (massage under water, Berthollet steam bath, and bath in suspension) Dax (hot mud), Bourbonne les-Bains and Bourbon l'Archambault (medicated baths and showers) In Italy, applications of hot mud at Acqui and at Abano, in England the medicated baths and pools at Bath and Harrogate, in Switzerland, Baden

Sometimes the different technics utilize water as such baths in a pool, showers and affusions, massage under water and sometimes steam the Berthollet steam baths at Aix sometimes vegeto-mineral muds for local or general applications. In a general way the technics employed in Ankylosing Spondylitis should be sedative percussion jets, violent massage, and brutal variations in temperature must be avoided

The modes of action have not been elucidated Heat is the essential element of this kind of treatment, but it is probably not the only one the chemical composition of the water its state of ionization, the presence of rare gases, and the radioactivity of the water, of which we have seen the favorable effects in the treatment of Ankylosing Spondylitis, also must be taken into account

The indications vary according to the stage of evolution of the disease, but hot spring treatment should be avoided during bouts of pain and in very weak patients When these methods are employed with discrimination, they bring to the patients an attenuation of the painful symptoms and an improvement in general condition and after orthopedic or surgical operations, they permit an accelerated re-education of the patient

ORTHOPEDIC CORRECTION

We have described the methods employed to prevent deformities. But because these are often neglected we see patients who, although they have received many kinds of internal medication present, because of failure to follow orthopedic precautions, deformities

which must be corrected A stiff or ankylosed joint, but in a position of function, allows the patient a satisfactory degree of activity On the contrary, when the same loss of mobility is accompanied by joints that are fixed in a faulty position, the patient becomes an invalid or a semi invalid

Long practice has made it possible for us to perfect different methods and to set forth the indications for them

The purpose of these methods is

1) To replace the affected joints in a position of function

2) To recover a certain degree of mobility when this is possible

3) To diminish, by improving the static condition of the joints, the excessive work done by these joints and by neighboring articulations, and thus to act on the inflammatory process itself

Correction of Different Deformities (According to Their Situation)

Spine

As far as the spine is concerned, we usually have to correct a dorsal or a dorsolumbar kyphosis.

Method of Straightening by means of Swaim's simple Orthopedic Appliances (210) The patient must lie on a "hard bed" all day for two three, or four weeks. He must stretch out on his back. When the patient has very marked deformities, the head should remain separated from the plane of the bed and, since this posture causes pain, a soft cushion should be placed under the head and the thickness of this cushion is gradually diminished A small pillow may be placed under the lumbar region in order to favor lordosis

In cases with marked or old deformity, straightening of the spine will be facilitated by means of bags containing from 1 5 to 2 0 kg of sand, which should be placed one over each knee, over the shoulders and, in case the head projects forward, over the forehead With the sand bags in place the patient remains in this position four or five hours a day, in two or more periods

Twice a day, and for periods of from 20 to 30 minutes, the cushion under the head should be removed and the patient should then place his hands under the nape of the neck with the elbows as near



Figure 143 Plaster corset with anterior support (Swaim type) (Photograph by J J Herbert)

as possible to the plane of the bed, so as fully to expand the thoracic cage.

When the patient has become accustomed to this horizontal position another pillow should be placed under the back, opposite the most prominent part of the kyphosis. Another expedient that may be useful in straightening the spine consists, when this is possible in having the patient lie flat on his belly with sand bags over the dorsal region, for periods varying from half an hour to an hour

During this period of straightening the patient is given either Aspirin or Dolosal (or better Butazolidine) which makes the straightening bearable, or Decontractyl which, by eliminating muscular contracture facilitates the straightening in a hot spring



Figure 144 Removable corset of plastic material, having the same shape as the Swaim plaster corset (manufactured by Sutter) a. Front view of the corset. b Back view of the corset. (Photographs by Muleau)

environment the contracture is also diminished by hot baths, either in a pool or with the so-called submarine spray

When this method is well applied, surprising results are obtained as far as the degree of straightening is concerned The rate at which straightening takes place is variable it depends on the degree of kyphosis and especially on the extent of the fibrous or bony nature of the stiffness or vertebral ankylosis. Often very rapid straightening is obtained the head comes in contact with the plane of the bed in from 10 to 13 days, in other cases a slightly longer period is necessary Bi weekly measurement of the head-to-wall distance permits one to determine when maximum straightening has been obtained, but in all cases the patient is kept in bed during a minimal period of *two or three weeks*

When the patient is completely or partly straightened, he must be maintained in this position by means of a Swaim corset (Fig 143), first in plaster bandages, and later in a removable plastic material (Fig 144) It is useless to undertake to straighten a kyphotic spine on a hard bed if when the spine has been straightened, it is not main

tained in this position by means of a plaster corset, because then the gain is rapidly lost

Swaim Corset (Maintenance Corset) (211) After long years of experience with the orthopedic treatment of Ankylosing Spondylitis, L T Swaim succeeded in perfecting a type of corset and in demonstrating its advantages over other types that had been used previously (212) In France, these advantages have been largely confirmed by J Forestier

This corset consists of a circular plaster cast which extends, anteriorly, from the pubic symphysis to the suprasternal notch, and posteriorly from the middle part of the buttocks to a line joining the points of the two scapulae. Thus the corset is strongly supported on the pelvis, compresses the abdomen, and keeps the dorsal spine from moving forward the thorax resting against the large plaster breast plate. But it also leaves free the upper part of the back The corset prevents the trunk from inclining forward and, besides, the thorax butts against the sternal support at each inspiration and is thrown back by it, thus the kyphosis is gradually corrected Moreover, thoracic respiration is favored by the limitation of abdominal respiration imposed by the corset.

Construction Technic (212) For six or seven days before the corset is applied, the skin should be cleansed daily with alcohol in order to harden it.

The corset can be applied with the patient either in vertical position and supported or not supported by a Sayre collar or, which is greatly preferable, in horizontal position on an orthopedic table When the patient is in the vertical position, he should hold himself as straight as possible with the arms slightly separated from the thorax when in the horizontal position, the lower extremities should be well extended

The iliac crests, the sacral region, and the prominent points of the spine should be protected with padding

The shape and limits of the plaster cast have been described To it may be added a lateral crutch to prevent aggravation of scoliosis. Plaster bandage permits better modeling than splints prepared in advance

While the plaster is still not completely dry, the anterior part of its lower border is cut away, so as to make sure that the thighs can flex at a right angle and that the patient can sit comfortably The

edges are "broken" throughout their extent so that they may not injure the patient's skin

Chin Support (Herbert) (104) In cases in which the kyphosis starts high, that is, above D-6, a chin support may be added to the Swaim corset, this should be fixed to the anterior-superior part of the corset and, by boxing in the mandible, limits the fall of the head According to the author, repeated opening of the mouth for eating or for conversation, with the mandible fixed, throws back the head and helps to straighten the spine

Except in a few cases, *the Swaim corset is well tolerated* and may be left in place Sometimes, because of abdominal compression, the patient may have difficulty in breathing after meals during the first few days, sedatives may then be given and food may be taken "on the instalment plan" Forty-eight hours after the cast has been applied, the discomfort has often disappeared and costal respiration has become easier and deeper

Muscular tension of the hips, as well as tension of the muscles at the nape of the neck, diminishes

The plaster cast, as described, remains in place for a month

When straightening is obtained, the sternal breast plate moves back from the anterior support and felt is slipped between the thorax and the plaster in order to maintain the anterior support After a month, when a considerable degree of straightening has occurred, a fresh cast should be applied to fit the improved posture When the kyphosis has been corrected to the maximum degree, *a removable corset* of plastic material should be provided, this should have the same shape and the same dimensions as the plaster corset (Fig 144)

During the weeks required for the manufacture of a plastic corset, the patient should continue to wear the plaster corset so as not to be without support. But the plaster corset will have been opened longitudinally at the sides in order to remove it and to model the removable corset, but it is preferable to do this modeling before applying the last plaster cast, so that it would not have to be opened

At first the removable corset should be worn night and day Later it may be removed at night [1] After one or two years, the patient

[1] Musicians who play wind instruments are obliged to remove the corset when they play because the corset limits their abdominal respiration.

may gradually cease to wear it but he should form the habit of wearing it one day a week, to make certain that the corrected posture has been maintained

Results of Straightening The results obtained by these methods are often surprising Patients with very marked and even very old *kyphoses are largely straightened* even patients who present an extensive bony ankylosis may derive benefit from this treatment. Only cases with a "bamboo' spine or an ankylosis of the *interapophyseal articulations do not improve.*

Swaim (212) uses this method, not only as a means of correction, but also as a basic treatment of the disease We have found that *the process is less tenacious in patients who have been straightened and whose improved position has been maintained than in those who have not been willing to accept this kind of treatment*

Indications 1) In patients who have a tendency to become round-shouldered or to bend forward (accurate measurement of the occiput to-wall distance is the best way to determine this), straightening should be carried out by means of the hard bed, and then the corset is applied

2) This treatment should also be applied to patients with a distinct kyphosis but who still have a certain degree of mobility

3) To be left without a corset are those patients who preserve a good station which has been maintained by the practice of good orthopedic hygiene, and those who have very extensive bony ankylosis

In patients with a good posture, but who suffer from pain, the corset is not used as a basic treatment, because the pain can be relieved by spinal treatment with roentgen rays or with radioactive agents

4) When the patient has a pronounced kyphosis that cannot be reduced by *simple orthopedic treatment, surgical treatment* (spinal osteotomy) should be considered

Hips

The hips are in large measure responsible for the attitude of the trunk in flexion (Occiput to-wall distance, p 16)

The Hard Bed which straightens the spine also corrects the flexed attitude of the hips. In certain patients rest in the ventral

position, with the head extended beyond the bed and with a sand bag over the pelvis, should be prescribed

Continuous Extension When very painful involvement of the hips is present, with or without flexion, continuous extension may be achieved with simple means linen bands surrounding the ankle, a cord, a pulley, and a weight of 2 or 3 kg Traction should be maintained for a month or a month and a half, often the pain diminishes about the tenth or fifteenth day, then exercises in active mobilization should very prudently be carried out.

Replacement of a Deformed Hip into a Functional Position This procedure, which is advocated by one of us (J Forestier) for osteoarthritis of the hip joint, has sometimes been employed in Ankylosing Spondylitis It may be advised only in cases with unilateral involvement and without bony ankylosis, when ankylosis is present, only a surgical operation will permit straightening

Knees

Stiffness of the knees in an attitude of flexion makes walking difficult or impossible, while at the same time it maintains the deformity of the hip in flexion (reciprocally the deformity of the hip maintains that of the knee), therefore, it must be corrected

When the deformity is minimal, it may be improved by rest on a hard bed Plaster shells for rest, combined with daily exercises, are more effective After extension has increased about 10°, a fresh shell should be applied in the new position, the successive application, at more or less long intervals of new plaster shells in better position may lead to complete correction and may thus constitute an excellent means which we often use To us continuous traction has not seemed to be effective when the knees were involved

Forced straightening of the knee under anesthesia, has very few indications in Ankylosing Spondylitis, because of the nearly constant association of hip involvement more marked than that of the knee, and because of the ankylosing tendency of peripheral involvement in Ankylosing Spondylitis, which is much more pronounced than in rheumatoid arthritis

As *the technics of straightening* have just been set forth *they are very different for the spine and for the extremities* it may be interesting to explain the results and the indications of these technics.

Kyphosis develops in a passive manner since the lumbodorsal muscles no longer exert their normal tension, the spine inclines forward while *loss of extension of the knees for example, is an active phenomenon* which is due to contraction of the ischio-femoral muscles. On the other hand, *the functional position of the spine is a position intermediate* between flexion and extension while *for the knees and to a certain extent for the hips also,*[1] *it is an extreme position* (complete extension).

The simple action of weight, which is exerted when the patient is lying on a 'hard bed' succeeds in gradually correcting the kyphosis when the corset is applied in a standing position, it will only maintain the correction of the spine. On the contrary when the knees are flexed, they do not find a position of rest, even when the patient is lying down. It is necessary first to suppress muscular spasm by means of plaster shells in the position in which the lower extremities happen to be (straightening by orthopedic maneuvers such as continuous traction will only serve to increase the muscle spasm). In spite of successive gains, it is often difficult to obtain complete extension and thus a certain degree of deformity persists. Therefore, the shells must be applied very early, before the development of any marked deformity.

SURGICAL OPERATIONS

These may be divided into two groups

1) Those that are based on pathogenic hypotheses and the object of which is to modify the evolution of the process

2) Those that aim to correct or to repair articular deformities or ankyloses resulting from the disease

Operations Aiming to Modify the Evolution of the Process

Parathyroidectomy

Oppel (148) thinking that the stiffness in Ankylosing Spondylitis was a manifestation of hyper parathyroidism, as confirmed by hypercalcemia, performed parathyroidectomy in 19 cases of 'Chronic rheumatism'. Since then this operation has been per

[1] The functional position of the wrist and ankle is an intermediate position but these joints have little stability. Therefore the wearing of plaster shells is necessary to maintain a good position

formed especially in a large number of cases of Ankylosing Spondylitis. The results have been discordant, aside from a few spectacular and more or less persistent improvements, practically only failures have been recorded Lièvre (140), who grouped the results of 32 operations, concluded that, at the present time, we do not have the elements necessary to judge the value of this operation, and that a fresh investigation, with systematic examination of the patients before and after operation, should be undertaken to determine its value Out of 14 cases, in which the patients were operated on by different surgeons we have observed a single instance of improvement, and in four cases in which the patients were operated on according to our indications, three failures and one improvement (in this case a thymectomy was also performed), practically speaking, the course of the disease has not been modified by the operations, and this does not encourage us to advise this operation Moreover, Oppel's hypothesis has not been confirmed from a clinical or biologic point of view (the calcemia was normal)

Since the discovery of the action of Cortisone in inflammatory rheumatism, every means has been sought to stimulate the suprarenal cortex either directly or through the hypophysis. Basing themselves on the experimental action of denervation of the carotid sinus, Lièvre and Léger have tried in rheumatoid arthritis, skinning" the carotid sinus, with some results on the objective articular signs But besides the results being far from constant and persistent, this operation is not devoid of danger, cases of death have been reported by Coste and by Michotte

Ligation of the inferior thyroid artery, which is usually carried out during parathyroidectomy and which is close to the carotid glomus, may perhaps explain some of the rare, spectacular and transient cures that have followed parathyroidectomy

Orthopedic Surgery

The problem of bloody orthopedic operations arises essentially for

Very marked and fixed kyphoses

Ankylosis of the hips

and less often for

Ankylosis of the knees,

Equinus deformities of the foot,
Ankylosis of other joints of the extremities,
Temporo-maxillary ankylosis

Always one must bear in mind that surgical operations permit one to solve mechanical problems, but that they do not have any influence on the disease itself. Thus surgery must be regarded as an *adjuvant* in the treatment of inflammatory rheumatism, and *not as a basic treatment*.

Before deciding upon an operation the *following conditions must always be fulfilled*

Conditions Affecting the Disease

An accurate diagnosis must have been made, when the necessary elements for such a diagnosis are not available, the physician must abstain from surgical procedures and must follow the evolution of the disease.

The operation must be performed *during a quiescent period*. As signs of progression, it is especially important to consider pain, joint swelling, osteoporosis (when this is marked, it is always a contra indication), fever, and increase in sedimentation rate. During the first years of active disease, one should be even more strict in observing this rule,

Before operating on a given joint, the condition of neighboring joints should be determined, and the operation should be integrated into a *complete orthopedic plan* which must include all the problems raised by the patient's other involved joints.

Conditions Relating to the Patient

The patient's general and muscular condition should be satisfactory.

His morale should be good and he should really desire to resume an active life.

The most favorable age for these operations is between 20 and 50 years. Below 18 years, one must not fail to consider that the epiphyses have not completed their development. Above 50 years, the individual less often has the physical stamina required for these operations and for the subsequent re-education, especially when the patient has been an invalid for many years,

When several operations must be performed, the physical condition of the patient, his age, the number of operations, and the possible advantages to be gained must be evaluated, and the physician must decide whether the advantages to be gained justify the effort which the operations would require,

Before proceeding with an operation, the physician should make certain that the patient has the time and the means necessary for him to rest in a center where adequate functional re-education will be compulsory We have observed numbers of cases in which, after the patients had been operated on with a correct technic, they were replaced too soon in an unfavorable environment (familial or other), and in which ankylosis of the operated joint rapidly recurred because they had been deprived of this functional re-education

Choice of Surgeon and Surgical Center

The surgery of rheumatism is very delicate, it raises multiple problems, and requires of the surgeon many qualities For operations on these patients there should be available

A specialized and patient surgeon

A surgical center organized not only for operations, but also for the functional re-education which must follow them

As far as rheumatic surgery is concerned, improvisation and haste lead only to disaster The collaboration of the rheumatologist and the surgeon is essential

The Spine. When the patient presents a very marked kyphosis, that is, one that makes his life difficult and that cannot be reduced by rest on a hard bed, and when this has continued a long time, *surgical straightening by spinal osteotomy* should be considered

The first spinal osteotomy was performed by Smith Petersen in March, 1941 (202)

This operation consisted of a resection from the spinous processes of the yellow ligament (ligamentum flavum) and of a partial resection of the apophyseal articular masses. This resection bore on two or three vertebrae in the lumbar region the level chosen was that in which the syndesmophytes, according to the roentgenographic films, were the least pronounced After this osteotomy, the patient was straightened on the operating table by raising the feet and head

Once the kyphosis had been corrected, small bone grafts from the resected spinous processes were inserted, and a large plaster cast was applied

In 1946 La Chapelle (133) reported a case of straightening a kyphosis in Ankylosing Spondylitis by a single stage posterior osteotomy (L 2 to L-3) at a second-stage operation, this was completed by an osteotomy of the anterior vertebral arc at the level of the disc, made through a large anterior incision below the umbilicus. The kyphosis was corrected on the operating table the small grafts were inserted and a large plaster cast was applied

Since 1947, J J Herbert has performed 14 osteotomies (106)[1] with a technic similar to that of La Chapelle In a first operation he sections the posterior arc by a trapezoidal resection of the spinous processes, the laminae, and the posterior wall of the foramina The patient's deformity is not corrected on the operating table but, after the operation, he is put to bed without a plaster cast but with cushions under the head to prevent an abrupt straightening When during the next few days, the patient gradually straightens, a plaster corset of the Swaim type is then applied and he may resume walking as early as the fifteenth day When the patient does not straighten after 10 or 15 days, an anterior osteotomy is performed as a second-stage operation

The first approach is through a left horizontal incision, beginning the width of three fingers outside of the navel and extending to the lumbosacral mass thus good access to the vertebral bodies and discs is provided Then the surgeon proceeds to resect a disc or only the anterior and lateral ligaments. The patient is then straightened on the table and a corset of the Swaim type is applied

Of the 14 patients operated on by J J Herbert, five could be straightened by a simple posterior osteotomy but in nine others it was necessary to perform a two-stage operation posterior and anterior

In 10 cases the *level of the operation* was lumbar D 12 and L-1 in one case, D 11 and D 12 in two cases, and high dorsal in one case.

In his series of 14 cases J J Herbert noted only a few pains due to traction on nerves at the time of straightening these pains oc

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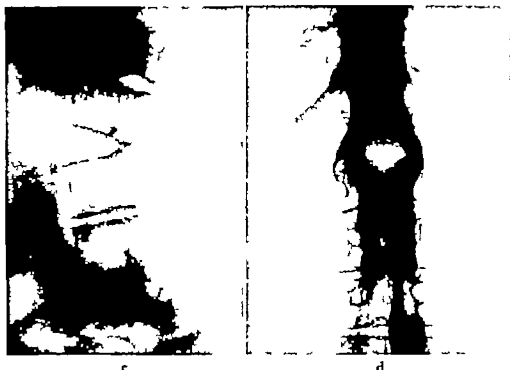
[1] In 1954 the number was over 40



Figures 145a and b (J J Herbert) Lumbar spine of a patient afflicted with Ankylosing Spondylitis a. Antero-posterior roentgenogram bony ankylosis of the interapophyseal joints, and ossification of the interspinous ligaments. b Antero-posterior roentgenogram the posterior arc L-1 and L-2 has been resected according to the technic of J J Herbert.

curred in the territory of the lumbar plexus, and especially in that of the femoral cutaneous nerve

The osteotomized spinal column undergoes rapid consolidation and during the year that follows the operation, a well organized and voluminous callus is noted in roentgenograms of the zone of the resected processes (Fig 145)



Figures 145c and d (J J Herbert) c. Lateral roentgenogram angular lordosis opposite L-1 and L-2 straightening was accomplished soon after the operation. d. Antero-posterior roentgenogram one year after the operation voluminous bony callus replacing the two interapophyseal joints L-1 and L-2.

RESULTS The straightenings obtained by *lumbar or dorsolumbar osteotomies* have been satisfactory they have permitted the patients to live under less difficult conditions they can look farther away their respiration being easier, their general condition has improved pain in the nape of the neck and in the thighs, which was due to muscular tension, disappeared after the spinal station had improved

The danger of recurring kyphosis is slight if the patient is faithful in resting on a hard plane and wears his corset for a few years. In a few cases, in which these two principles had been neglected by the patient, we have seen the recurrence of a certain degree of kyphosis, thus diminishing the benefits obtained from the operation

Osteotomies performed in the dorsal region by different surgeons (202, 106) have given negative or very meager results We believe these failures are related to ankylosis in a single block of all the elements of the thoracic cage, which prevents any straightening

of the dorsal segment The two last floating ribs, which are less firmly attached than the rest of the thoracic cage, nevertheless permit straightening at the level of D-11, D 12, and L-1

INDICATIONS The operation is indicated in cases of marked kyphosis that is not reducible by simple orthopedic treatment and with a lumbar or dorsolumbar starting point The highest level at which the operation can be performed is D-11 and D 12, where the ribs are still free Operations on the dorsal region, with the technics now available are not advised, this restricts the indication for osteotomy because of the frequency of kyphosis originating in the dorsal region

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Figures 145c and d (J J Herbert) c. Lateral roentgenogram angular lordosis opposite L-1 and L-2 straightening was accomplished soon after the operation. d Antero-posterior roentgenogram one year after the operation voluminous bony callus replacing the two interapophyseal joints L-1 and L-2.

RESULTS The straightenings obtained by *lumbar or dorsolumbar osteotomies* have been satisfactory they have permitted the patients to live under less difficult conditions, they can look farther away, their respiration being easier their general condition has improved, pain in the nape of the neck and in the thighs, which was due to muscular tension disappeared after the spinal station had improved

The danger of recurring kyphosis is slight if the patient is faithful in resting on a hard plane and wears his corset for a few years. In a few cases, in which these two principles had been neglected by the patient, we have seen the recurrence of a certain degree of kyphosis, thus diminishing the benefits obtained from the operation

Osteotomies performed in the dorsal region by different surgeons (202, 106) have given negative or very meager results We believe these failures are related to ankylosis in a single block of all the elements of the thoracic cage, which prevents any straightening

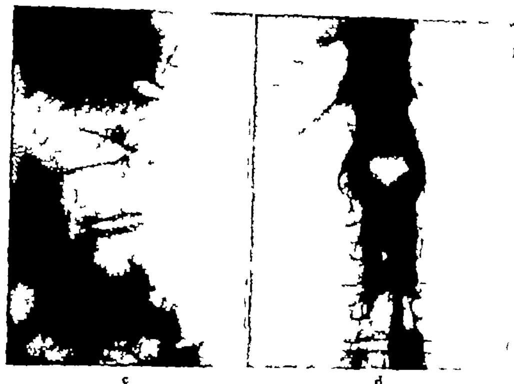
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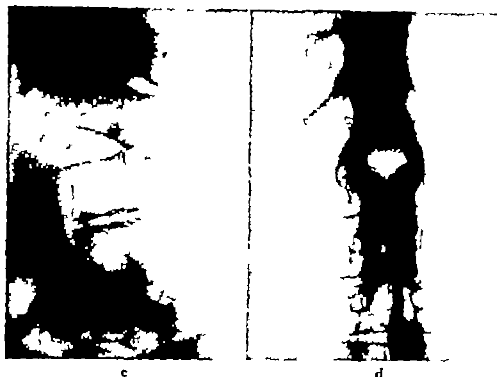
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Spondylitis, but especially for the hips in which ankylosis occurs frequently and early, given its usual bilateral distribution, this involvement is very serious from a functional point of view

At first the surgeons used the same technics as for the osteoarthritides or for rheumatoid arthritis simple arthroplasties with resection, with or without the interposition of fascia, have been rapidly abandoned because the ankylosis recurred very rapidly Two technics are now established arthroplasty, with interposition of a Vitallium cup (Smith Petersen), and resection of the femoral head and its replacement by a head made of acrylic resin (Judet) [1] The approach may first be either anterior, as favored by Smith Petersen (201), or external with section of the greater trochanter which, at the end of the operation, is restored to its place

POST OPERATIVE CARE The patient's hip remains immobilized for 15 days, in slight abduction and internal rotation, the joint being immobilized by means of continuous extension, then passive and active mobilization are begun, five or six weeks after the operation, the patient may begin to walk, using crutches Smith Petersen strongly insists on the fact that, when a cupula is inserted, the patient must not put his full weight on the hips until after the 6th month in order to permit the reconstitution of a sort of articular cartilage When a pool of warm water is available the patient may be required to take exercises in water as early as the twentieth or the twenty fifth day

RESULTS The results are much less favorable than those obtained in cases of osteoarthritis and even than those obtained in rheumatoid arthritis

Smith Petersen followed 20 cases from one to six years, and these gave

Eight unilateral arthroplasties 12% of good results, 38% of reasonably good results and 50% proved to be failures

Twelve bilateral arthroplasties 33% of good results 17% of reasonably good results and 50% proved to be failures

Other reports (134) are more favorable, but they do not separate cases of spondylitis and rheumatoid arthritis Of the few cases that

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[1] J Gosset uses prosthetic specimens including a head, a neck and a stem penetrating into the medulla of the femoral diaphysis.

were operated on on the basis of our own indications nearly all ended in failure

According to Smith Petersen, one of the causes of failure[1] may have been a subluxation of the cup due to proliferation at the base, this proliferation is related to the stage of the disease and to the very slight pressure exerted by the atrophied gluteal muscles on the bottom of the acetabulum Smith Petersen suggests that the operation should be performed in two stages the first stage would permit a slight mobility that would be susceptible to a certain degree of muscular recuperation, and later the second stage would consist in restoring the cavity of the acetabulum this second stage would then yield better results

But it is especially the bony proliferation which, by reproducing itself around the cupula surrounds the head and reduces the mobility of the hip and the ankylosis rapidly recurs

The results obtained with Judet s technic appear to be more encouraging, re-education is easier and requires less time To judge the value of this technic it will be necessary to wait a few years. But our impression is that the results are better in the form of disease with bony ankylosis than in the form with active arthritis

At the present time considering these results, *operation should be undertaken only in cases presenting a marked bilateral ankylosis or stiffness that interferes considerably with the patient s life* Perhaps the combination of hormone treatment and surgery may serve to improve the results and widen the indications.

The Knees *Involvement of the knees occurs frequently and may raise different problems*

1) In the case of ankylosis of *a straight knee* or of one with a loss of extension not exceeding 15% or 20%, with good mobility of the homologous hip and allowing the patient to walk without too much difficulty the question of surgery does not arise

2) When *both knees are ankylosed in a straight position*, the question of arthroplasty for these joints may arise Putti s technic, with the insertion of *fascia lata* is no longer advised (106) ankylosis often recurs or the patient has an unstable joint.

[1] *Another cause of failure is absorption of the stump of the head within the cupula the head may come out of this and the cupula may sink into the neck Surgeons think this is due to an inadequate resection of the diseased parts this is especially true for osteo-arthritis of the hip.*

by J J Herbert (106) The disadvantages of this operation are 1) Early recurrence of ankylosis To prevent this, mobilization should be started 10 or 15 days after operation, 2) Loss of joint stability, usually a minor disadvantage, 3) Lesions of the ulnar nerve that produce paralysis and a deformed hand which is very rebellious to treatment Nevertheless, this operation should be attempted in order to give the patient a certain degree of independence

The Temporo-Maxillary Joints (Analysis of Symptoms, p 154)

MANNER OF CONDUCTING THE TREATMENT

We have just analyzed the different elements that should be employed in treating Ankylosing Spondylitis. We will now explain how these elements should be combined in specific cases

Two problems should be solved

The treatment of the inflammatory process itself aiming to arrest the progress of the disease, or the basic treatment

The prevention and correction of deformities

The Basic Treatment

In choosing one of the elements that have been considered as acting on the progress of the disease (p 304), we take into account the following conditions

In painful cases with local or diffuse involvement of the spine, but without peripheral involvement, either treatment with roentgen rays, Thorium X, or Radon should be chosen and should be employed in the doses indicated,

In cases with marked peripheral involvement or in which the sedimentation rate is very high, gold salts should be selected When the patient has one or two very painful joints (which sometimes occurs in the hips), treatment with roentgen rays may be indicated Thorium X also may be very useful when other kinds of treatment have failed

Forms of Ankylosing Spondylitis with psoriasis should be treated

with Thorium X or gold salts with the precautions mentioned, or with salts of copper,

The latent forms, the ill-defined forms, and the advanced forms in which the disease is not very active, should not be treated with the basic methods of treatment.

The choice of one or another of these elements should be made, in principle, according to the rules that have been mentioned, but in Ankylosing Spondylitis there is no absolute indication for the use of one or another of these elements The choice will often be made according to the conditions under which the treatment may best be carried out in the country, for example, treatment with roentgen rays will be replaced by Thorium X The difficulty of giving intravenous injections may lead one to exclude Thorium X or Cupralene Intolerance of gold salts may cause these to be avoided

Courses of Thorium X or Radon may be given, one or the other, according to the possibilities

Courses of Treatment

All these elements will be employed in courses and in the doses indicated, the courses should be separated by *periods of rest* In cases in which the treatment selected should not bring about improvement, another method may be tried, but one should never judge the effectiveness of one of these methods after only a few injections or after a single course of treatment. When the treatment is changed, one should allow the period of rest indicated *Never should two of these methods of treatment be employed simultaneously*

Stopping the Treatment

Stopping the basic treatment is a very delicate problem the patient should be treated systematically as long as the disease shows signs of progression Many physicians confine themselves to relieving the pain but this is a grave error, one should consider all the signs that may reveal the progressive character of the disease these signs are

Spontaneous pain, even when this is slight

Swelling of joints,

Bouts of iritis,

Increase in deformity, for example, of kyphosis with an increase of a few centimeters in the occiput to-wall distance,

Increase in the rate of sedimentation

A sedimentation rate above the normal level (between 10 and 20 mm in 1 hr), which we call "residual," does not oblige one to pursue the treatment when other signs of progression are absent

As long as signs of progression continue, the patient should be treated and, after these signs have disappeared, it is wise to continue the treatment for one or two years with moderate and well spaced courses.

After the treatment has been stopped, it is necessary to warn the patient of the possibility of fresh attacks and to keep him under *periodic observation every six or 12 months* Recurrence of any of the symptoms should be the signal to resume courses of treatment.

Prescribing a basic treatment should include instructions on the patient s living conditions

When the patient's general condition is deficient, one of the adjuvant methods of treatment that have been indicated should be given between courses of basic treatment (calcium therapy, vitamin therapy), in a few cases in which the patient presents a general lassitude or anemia, blood transfusions may improve his general condition and facilitate the basic treatment.

Preventive Orthopedic Treatment

We have already emphasized the importance of this kind of treatment. Before explaining to the patient the rules of orthopedic hygiene (hard bed which should be ordered in every case plaster shells, etc.), a careful examination of the joints should be made

Already established deformities should be corrected by means of plaster shells, posture exercises, etc When a deformity must be corrected surgically, the operation should be considered in relation to the general treatment

TOTAL RESULTS

It is difficult to estimate the results of treatment for a disease with such a long evolution and for one that is so characterized by exacer

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References

1 ARCUSA S M and SANS L I Estadistica y comentarios de 153 casos de espondyloartritis anquilopoyetica. *Rev españ reum* *1* 216–221, 1945

2. D ARSONVAL, A. Applications médicales des émanations radioactives d'après les travaux de M G Vaugeois. *C R Acad Sci (Paris)* *191* 2101–2102 1932.

3 BARCELO, P and VILASECA-SABATER J M Espondilitis melitococicas clinica y radiologia. *Med clin* *3* 184 198 1944

4 BARCELO, P *Reumatismos vertebrales* Barcelona Salvat 1945

5 BARCELO P and VILASECA SABATER J M Espondilitis tifica. *Rev españ reum.,* *1* 64 1945.

6. BARR J S Relationship of intervertebral disk to back strain and peripheral pain (sciatica). *Surgery* *4* 1–12, 1938

7 BARSONY T and POLGAR F Osteitis condensans ilii ein bisher nicht beschriebenes Krankheitsbild *Fortschr a.d Geb d Röntgenstr* *37* 663–669 1928

8 BATALLA BOIXET E. and ROTES-QUEROL, J Sacroileitis unilateral como forma de comienzo de la espondiloartritis anquilopoyetica. *Rev españ reum.,* *2* 488–495 1948.

9 BATSON O V Function of the vertebral veins and their role in the spread of metastases. *Ann. Surg.,* *112* 138–149 1940

10 BECHTEREW W L V Steifigkeit der Wirbelsäule und ihre Verkrümmung als besondere Erkrankungsform. *Neurol C. B L.* *12* 426–434 1893

11 BECHTEREW W L V Von der Verwachsung oder Steifigkeit der Wirbelsäule. *Deutsche Ztschr Nervenh.,* *11* 327–337 1897

12. BECHTEREW W L V Ueber ankylosierende Entzündung der Wirbelsäule und der grossen Extremitärengelenken. *Deutsche Ztschr Nervenh.* *15* 37–44 1899

13. BECHTEREW W L V Neue Beobachtungen und pathologische-anatomische Untersuchungen über Steifigkeit der Wirbelsäule. *Deutsche Ztschr Nervenh.,* *15* 44–57 1899

14 BENNETT G A. Comparison of pathology of rheumatic fever and rheumatoid arthritis. *Ann. Int. Med* *19* 111–115 1943

15 BERENT F (quoted by Hare and Haggart [See Ref. 100]) Beiträge zur Pathologie der Kreuzdarmbeinfügen. *Arch. orthop u. Unfall-Chir* *32* 642–646 1933

16. BERGER (quoted by Schinz Baensch and Friedle [See Ref. 188]).

17 BERNSTEIN L. and BLOCH O J Cardiac complications in spondylarthritis ankylopoietica. *Acta med. scandinav.,* *135* 185–194 1949

18 BICKEL, W H and BARR, F S Cup arthroplasty of the hip. *J Bone & Joint Surg.,* *30-A* 647–656 1948

19 BOLAND E. W and PRESENT A J Rheumatoid spondylitis study of 100 cases, with special reference to diagnostic criteria. *J.A.M.A* *129* 843–849 1945

20 BOLAND E. W and SHEBESTA, E. M Rheumatoid spondylitis correlation of clinical and roentgenographic features. *Radiology* *47* 551–561, 1946.

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References

1 Arcila, S. M. and San J. Estadística y comentarios de 153 casos de espondiloartritis anquilopoyética. *Rev. españ. reum.* 1 21[illegible] 221 1945
2. d'Arsonval A. Applications médicales des émanations radioactives d'après les travaux de M. C. Vaureen. *C. R. Acad. Sc. (Paris)* 19[illegible] 2101 2102 1932.
3. Barcelo, P. and Vilaseca Sabater J. M. Espondilitis melitococicas clinica y radiologia. *Med. clin.* 3 1[illegible]4 19[illegible] 1944
4. Barcelo, P. *Reumatismo vertebral.* Barcelona Salvat 1945
5. Barcelo, P. and Vilaseca Sabater J. M. *Espondilitis tifica. Rev. españ. reum.,* 1 64 1945
6 Barr, J. S. Relationship of intervertebral disk to back strain and peripheral pain (sciatica). *Surgery* 4 1 17 193[illegible]
7 Barsony T. and Polgar F. Ostitis condensans ilii ein bisher nicht beschriebenes Krankheitsbild. *Fortschr. a. d. Geb. d. Röntgenstr.* 3[illegible] 663–667 1928
8. Batalla Borrell L. and Roura Oterino J. Sacroileitis unilateral como forma de comienzo de la espondiloartritis anquilopoyetica. *Rev. españ. reum.* 2 488–495 1948
9 Batson O. V. Function of the vertebral veins and their role in the spread of metastases. *Ann. Surg.* 11[illegible] 138 147 1940
10. Bechterew W. L. V. Steifigkeit der Wirbelsäule und ihre Verkrümmung als besondere Erkrankungsform. *Neurol. C. Bl. L.* 12 426–434 1893
11 Bechterew W. L. V. Von der Verwachsung oder Steifigkeit der Wirbelsäule. *Deutsche Ztschr. Nervenh.* 11 3[illegible] 337 1897
12. Bechterew W. L. V. Ueber ankylosierende Entzundung der Wirbelsäule und der grossen Extremitätengelenken. *Deutsche Ztschr. Nervenh.,* 15 37–44 1899
13. Bechterew W. L. V. Neue Beobachtungen und pathologische-anatomische Untersuchungen über Steifigkeit der Wirbelsäule. *Deutsche Ztschr. Nervenh.,* 15 44 57 1899
14. Bennett G. A. Comparison of pathology of rheumatic fever and rheumatoid arthritis. *Ann. Int. Med.* 19 111–113 1943
15. Berent F. (quoted by Hare and Haggart [See Ref. 100]), Beiträge zur Pathologie der Kreuzdarmbeinfügen. *Arch. orthop. u. Unfall-Chir.* 32 642–646 1933
16. Berger (quoted by Schinz Baensch and Friedle [See Ref. 188]).
17 Bernstein L. and Broch O. J. Cardiac complications in spondylarthritis ankylopoietica. *Acta med. scandinav.* 135 185–194 1949
18. Bickel, W. H. and Babb F. S.: Cup arthroplasty of the hip. *J. Bone & Joint Surg.,* 30-A 647–656 1948
19 Boland E. W. and Present A. J.: Rheumatoid spondylitis study of 100 cases with special reference to diagnostic criteria. *J.A.M.A.* 129 843–849 1945
20 Boland E. W. and Shebesta, E. M.: Rheumatoid spondylitis correlation of clinical and roentgenographic features. *Radiology* 47 551–561 1946

21 Boland E. W Headley N E. and Hench P S The cerebrospinal fluid in rheumatoid spondylitis. *Ann. Rheumat Dis* 7 196–199 1948
22. Boland E. W Rheumatoid spondylitis. In Comroe, B I *Arthritis and Allied Conditions* 4th Ed. Philadelphia, Lea & Febiger 1949 559–593
23 Borak, J Significance of the sacro-iliac findings in Marie-Strümpell's spondylitis. *Radiology* 47 128–141 1945
24 Borellini, A. Aspetti anatomo-patologici dello scheleto di due casi di spondiloartrite anchilopoietica. *Chir org movimento* 30 372–382, 1946
25 Bouwens Van der Boijen G Le rhumatisme psoriasique (Thesis). Paris, Lefrançois, 1939
26 Briçon P Exostoses, hyperostoses et synostoses multiples de la colonne vertébrale chez un chat. *Bull. Soc anat (Paris)* 9 478–482, 1884
27 Editor, British Medical Journal Muscle biopsy in rheumatoid arthritis. *Brit. M J* 1:815 1949
28 Brocher, J E. W Le diagnostic précoce de la spondylarthrite ankylosante. *Radiol. clin.,* 16 389–392, 1947
29 Brocher, J E. W and Forestier, J *La Spondylarthrite Ankylosante* Lausanne, Rouge, 1949
30 Buckley C. W Spondylitis deformans. *Brit M J.,* 1 1108–1112, 1931
31 Buckley C. W Spondylitis deformans its differential diagnosis and treatment by physical methods. *Proc Roy Soc Med.,* 26 105–110 1932.
32. Buckley C. W *Ankylosing spondylitis Reports on chronic rheumatic diseases* , London, Lewis, 1935 77
33 Buckley C. W Ankylosing spondylitis its etiology and pathology *Ann. Rheumat. Dis* 5 49–54 1945
34 Campbell, A. M G Ankylosing spondylitis. *Lancet* 252 406–408 1947
35 Campbell Golding F Spondylitis ankylopoietica. *Brit. J Surg.,* 25 484–500, 1936
36 Christin M La vitesse de sédimentation au cours de la spondylarthrite (Oral communication).
37 Clausen F and Kober, L. (Quoted by West, H F [See Ref. 218]). Ueber die Veranlagung zu Bechterewscher Krankheit und ihr Wesen. *Ztschr Konstlehre* 22 268–303 1938
38 Cohen A. Goldman J and Dubbs, A W Treatment of rheumatoid arthritis with 417 courses of gold analyses of 259 cases. *New England J Med* 233 199–203 1945
39 Coggeshall, H. C. Backache. In Comroe, B. I. *Arthritis and Allied Conditions* 4th ed. Philadelphia Lea & Febiger 1949 981–1061
40 Coggeshall, H C. The synovial fluid. In Comroe, B I *Arthritis and Allied Conditions* 4th ed. Philadelphia, Lea & Febiger 1949 47–54
41 Collins, D H. *The pathology of articular and spinal diseases* London, Arnold, 1949 313
42. Comroe, B. I. *Arthritis and Allied Conditions* 4th ed. Philadelphia, Lea & Febiger 1949
43 Connor, B. Sur la continuité de plusieurs os (Thesis). Rheims (France), 1691
44 Copeman W S C. *Textbook of the Rheumatic Diseases* Edinburgh Livingstone, 1948 306.
45 Coriat P Anatomie radiologique de l'articulation sacroiliaque (Thesis). Algiers, 1935

46 Coste, F and Forestier J Remarques sur le rhumatisme psoriasique. Rev rhum., 2.554–558 1935
47 Costé, F., Forestier, J and Mande, R. Danger de certaines interventions chirurgicales au cours des polyarthrites chroniques évolutives. Presse méd 45 729–730 1937
48 Coste, F Les médications radio-actives dans les affections rhumatismales. Rev rhum., 4 759 820 1937
49 Curtis, A S and Pollard H M Felty's syndrome Its several features including tissue changes compared with other forms of rheumatoid arthritis. Ann. Int Med 13.2265–2284 1940
50 Davison R. A., Koets P and Kuzell, W C. Excretion of 17-ketosteroids in ankylosing spondylarthritis and in rheumatoid arthritis preliminary report. J Clin. Endocrinol 7.201–204 1947
51 Dechaume, M Nécroses des maxillaires consécutives à la curie thérapie interne. Mém Acad chir 62 1119–1125 1936
52. Desmarais M H L. The phosphatase activity in spondylitis ankylopoietica. Ann Rheumat Dis., 7 105–107 1948
53. Desmarais, M. H. L. The neutral 17-ketosteroids in rheumatoid arthritis and spondylitis. Ann. Rheumat Dis., 8.296–298 1949
54 Edström G Is spondylarthritis ankylopoietica an independent disease or a rheumatic syndrome? Acta med scandinav 104.396–413 1940
55 Ehrlich (Quoted by Overgaard, K. [See Ref. 153]) Die sog Bechterewsche Krankheit. Arbeit und gesundheit Schriftenreihe. Ztschr Rocherbeitsblatt No. 15 1930
56. Eldaroff N La spondylose rhizomélique est une entité morbide spéciale. Nouv Iconogr Salpêtrière 24 121–139 236–255 1911
57 Fagge, C. H. Cases illustrating some remote effects of spinal deformities. Guy's Hosp Rep 19 189–212, 1874
58 Fischer, A. and Wurtz, O Klinik der Spondylarthritis ankylopoetica. Mitt Geb Med. u. Chir 42.586–606 1930–1932.
59 Fombeure, G and Lacapère, J Bio-microscopie et traitement des iridocyclites diffuses dites rhumatismales. Arch Hosp., 7.295–312, 1933
160 Forestier H. De la spondylose rhumatismale ou rhumatisme vertébral chronique sa forme pseudo-névralgique. Arch. gén. méd. (Nouv Sér) 129–168 1901
61 Forestier, J Le trou de conjugaison vertébral et l'espace épidural. Etude anatomique et clinique. Essai de phathologie de l'espace épidural (Thesis No. 77). Paris, Jouve, 1922.
62. Forestier, J Le diagnostic des rhumatismes vertébraux d'après les méthodes modernes d'examen. Presse therm. clim., 72.268–275 1931
63 Forestier J and Robert P Ostéophytes et syndesmophytes. Gaz. méd. France (Suppl. Radiol.), 196–202, 1934
64 Forestier, J Nature de la spondylose rhizomélique, physiopathologie et déductions thérapeutiques. Rev rhum., 2 472, 1935
65 Forestier, J Collez R. and Robert P Etudes radiologiques de pièces sèches de rhumatisme vertébral. Bull et mém Soc radiol méd France 23:612–624 1935
66 Forestier, J L arthrite sèche sacro-iliaque existe-t-elle? Arch. rhum 1(3).28–29 1936.

67 FORESTIER, J Le diagnostic des débuts de la spondylose rhizomélique. *Concours méd.*, No. 13 1936

68 FORESTIER, J and CERTONCINY A. Les déformations vicieuses chez les rhumatisants chroniques leur mécanisme et leur prévention. *Gaz. méd. France* 45:645 723 1938

69 FORESTIER, J and CERTONCINY A. Le rôle de l'infection gonococcique dans l étiologie de la spondylarthrite ankylosante. *Rev rhum.* 5:969–978 1938

70 FORESTIER J and METZGER, J La clef du diagnostic précoce de la spondylarthrite est dans la radiographie des articulations sacro-iliaques. *Presse méd* 47 1247–1249 1939

71 FORESTIER, J The importance of sacro-iliac changes in early diagnosis of ankylosing spondylarthritis Marie-Strümpell-Bechterew disease, *Radiology* 33:389–402, 1939

72. FORESTIER, J Recherches anatomo-radiologiques sur la structure des ossifications péri-vertébrales. *Rev rhum.*, 8:565–588 1941

73 FORESTIER J and CERTONCINY A Les attitudes vicieuses et les déformations articulaires chez les rhumatisants. *Arch. rhum* 5 73–77 1943

74 FORESTIER, J FRANÇON F and HERBERT J J Attitude en flexion extrême des genoux traitée par désinsertion capsulaire et extension continue. *Mém. Acad. chir (Paris)*, 69 148–149 1943

75 FORESTIER, J., FRANÇON F and HERBERT J J Quelques résultats du redressement orthopédique des spondylarthrites. *Rev rhum.*, 13 182–185 1946.

76. FORESTIER, J and CERTONCINY A. Le traitement des rhumatismes chroniques par les sels organiques de cuivre. *Presse méd.*, 54.584–885 1946.

77 FORESTIER, J., JACQUELINE, F and LENOIR, S. La cuprothérapie intramusculaire dans les rhumatismes chroniques inflammatoires. *Presse méd* 56.351–352, 1948

78 FORESTIER, J., JACQUELINE, F and ROTES, J Etudes statistiques sur les symptômes de début de la spondylarthrite ankylosante. *Rev rhum.*, 16.218–225 1949

79 FORESTIER, J HERBERT J J JACQUELINE, F and ROTES-QUEROL, J Aspects pseudo-chirurgicaux de début de la spondylarthrite ankylosante. Importance de la radiographie des sacroiliaques. *Rev orthop.*, 35 499–516 1949

80 FORESTIER, J ROTES-QUEROL, J and JACQUELINE, F Les articulations sacro-iliaques dans la spondylarthrite ankylosante. Radiologie des articulations sacro-iliaques normales. *Rev rhum* 17 407–448 1950

81 FORESTIER, J and ROTES-QUEROL, J Hyperostose ankylosante vertébrale sénile. *Rev rhum.*, 17.525–534 1950

82. FRAENKEL, E. Ueber chronische ankylosierende Wirbelsaülenversteifung *Fortschr a. d Geb d. Röntgenstr* 7·62–90 1903–1904

83 FRAENKEL, E. Ueber chronische ankylosierende Wirbelsaülenversteifung *Fortschr Geb Röntgenstrahlen*, 11 171–195 1907

84 FRANCESCHETTI, A. and BROCHER, J E. W Die Iridocyclitis "rheumatica" vom Standpunkt der Ophtalmologie und Rheumaforschung *Schweiz med Wchnschr.*, 74.299 1944

85 FRANCESCHETTI A. Les affections oculaires d'origine rhumatismale. *Ophtalmologica*, 111.242–269 1946

86. FRANÇON F. *Leçons Cliniques de Rhumatologie Pratique* Paris, Vigot, 1947

87 FRANÇON F and PELLAS, M Que peut-on attendre de la phosphatasémie et de la phosphorémie minérale pour le diagnostic de la spondylose rhizomélique? *Arch. rhum* 5 195–198 1944

88 FREUND H A. STEINER, G LEICHTENTRITT B., and PRICE A E. Peripheral nerves in chronic atrophic arthritis. *Am. J Path* 18:865–893 1942

89 FREUND, H. A. A contribution to the pathogenesis of spondylitis ankylopoietica. *Edinburgh M J* 49·91–109 1942.

90 FREUND, H A. STEINER, G LEICHTENTRITT B. and PRICE A. E. Nodular polymyositis in rheumatoid arthritis. *Science* 101.202–203 1945

91 FREYBERG R. H. Roentgen therapy for rheumatic diseases. *M Clin. North America* 30·603–615 1946

92. GEILINGER W Beitrag zur Lehre von der ankylosierenden Spondylitis mit besonderer Berücksichtigung ihrer Beziehungen zur Spondylitis deformans. *Ztschr f Orthop Chir*, 38 185–269 1918

93. GIBSON H J Special pathology of the rheumatic diseases. In Copeman W S C., Ed. *Textbook of the Rheumatic Diseases* Edinburgh, Livingstone, 1948 405

94 GIBSON H. J DESMARAIS, M. H L., and KERSLEY G D Muscle histology in rheumatic and control cases study of 119 biopsy specimens. *Ann. Rheumat. Dis* 7 132–141 1948

95 GOLDFAIN E. Chronic brucellosal type of ankylosing spondylitis. *J Lab & Clin. Med.*, 28 1226–1231 1943

96 GRABER DUVERNAY J Spondylarthrite ankylosante (spondylose rhizomélique) d'origine traumatique. *J méd Lyon*, 321–325 1948.

97 GRABER DUVERNAY J Les arthrites chroniques sacro-iliaques non tuberculeuses. *Arch. rhum.* 1(3).3–19 1936.

98 GÜNTZ E. Beitrag zur pathologischen Anatomie der Spondylarthritis ankylopoetica. *Fortschr a.d Geb d Röntgenstr* 47 683–693, 1933

99 HAPPEL, P Das Sakroiliakalgelenk im Röntgenbild. I Mitteilung Die alters und Geschlechtsverschiedenheiten des Sakroiliakalgelenks im Röntgenbild. *Arch. orthop u. Unfall-Chir.*, 20.576–596, 1922.

100 HARE, H. F and HAGGART G E. Osteitis condensans ilii. *J.A.M.A* 128 723–727 1945

101 HARVIER P TRAVERSE, M DE, DEPRAITÈRE, R. and COQUELET M L. Equilibre protidique du sérum sanguin chez les rhumatisants. *Ann. Inst. Hydrol (Paris)*, 20·96–127 1949

102. HENCH, P S., KENDALL, E. C., SLOCUMB, C. H. and POLLEY H F The effect of a hormone of the adrenal cortex 17 hydroxy 11-dehydrocorticosterone (Compound E) and of pituitary adrenocorticotropic hormone on rheumatoid arthritis preliminary report. *Proc Staff Meet. Mayo Clin.* 24 181–197 1949 and *Ann. Rheumat. Dis.*, 8·97–100 1949

103. HENCH, P S. KENDALL, E C. SLOCUMB, C. H. and POLLEY H F Effects of cortisone acetate and pituitary ACTH on rheumatoid arthritis, rheumatic fever and certain other conditions study in clinical physiology *Arch. Int Med.*, 85 545–666, 1950

104 HERBERT J J Comment remédier à la chute de la tête dans les spondyloses. *Arch. rhum.* 5.205–206 1945

105. HERBERT J J Vertebral osteotomy technique, indications and results. *J Bone & Joint Surg* 30·680–689 1948

106. HERBERT J J *Chirurgie et Orthopédie du Rhumatisme* Paris, Masson, 1950
107 HERMAMAN JOHNSON F and LAW A. W *Ankylosing Spondylitis A Practical Guide to Its Diagnosis and Treatment.* London, Butterworth, 1949
108 HORWITZ M. T and SMITH R. M. Anatomical pathological and roentgenological study of the intervertebral joints, of the lumbar spine and of the sacroiliac joints. *Am. J Roentgenol., 43* 173–186 1940
109 HUC, G Syndrome du trapèze. Cyphose. *Traité de chirurgie orthopédique 11* 1560 Paris, Masson, 1937
110 INGELRANS P and OBERTHÜR, M Les arthrites chroniques sacro-iliaques non tuberculeuses). *Rev orthop 35*.281–318 1949
111 ISAACSON A. S. and WHITEHOUSE, W M. Spontaneous sacro-iliac obliteration in patients with tuberculosis. *J Bone & Joint Surg 31 A*.306–311 1949
112. JACKSON R. H Chronic sacro-iliac sprain with attendant sciatica. *Am J Surg 24* 456–477 1934
113 JACQUELINE, F ARLET J and LAPORTE, C. La coxite densifiante à évolution bénigne. *Rev rhum 17* 114–120 1950
114 KAHLMETER, G The roentgen treatment of arthritis. *Brit. J Actinother 5*·93 1930
115 KAHLMETER, G *Manuel Pratique de Rhumatologie Etiologie Diagnostic et Traitement* Paris, Expansion Scientifique Française, 1949 136
116. KELLGREN The anatomy and physiology of pain In Copeman W S C., Ed. *Textbook of the Rheumatic Diseases* Edinburgh, Livingstone 1948 22.
117 KIENBÖCK, R. Bechterewsche Wirbelsaülenversteifung *Wien. Arch. inn. Med 32*.311–316 1938
118 KÖHLER A. Les limites de l'image normale et les débuts de l'image pathologique. *Roentgenologie* Paris, Delachaux et Niestlé 1936 315 318 432.
119 KOHEN J Contribution à l'étude de la spondylose rhizomélique traumatique (Thesis No. 315) Paris, 1935
120 KNAGGS, R. L. A report on the Strangeways Collection of Rheumatoid Joints in the Museum of the Royal College of Surgeons. *Brit. J Surg., 20* 113 309 425–443 1932–1933
121 KUIPERS, R. K. W Orthopaedic treatment of ankylosing spondylarthritis. *Lancet, 2* 774–775 1945
122. KREBS W Klinik der sogenannten rheumatischen Wirbelsaülenerkrankungen. *Rheumaprobleme 2* 103–110 Leipzig Thieme, 1931
123 KREBS, W and VOHTZ O Entstehung und Verlauf der Spondylitis ankylopoietica (Bechterew). *Deutsche med Wchnschr 60* 100–102, 1934
124 KREBS, W Beitrag zu den Beziehungen des Periosts und fibrösen Gewebes zu den rheumatischen Erkrankungen. *Rheumaprobleme 3*.36 Leipzig Thieme, 1934
125 KREBS, W and WURM, H Die Bechterewsche Krankheit. Die pathologische Anatomie der Bechterewschen Krankheit. *Rheumatismus 3* 1–94 1938
126 KREMER AND WIESE *Tuberculosis Osteoarticular* Barcelona Tabor
127 KÜMMEL, H Uber die traumatischen Erkrankungen der Wirbelsaülle. *Deutsche med Wchnschr 21* 180–181 1895
128 LABORDE, S. Sur les dangers des substances radio-actives introduites dans l or ganisme. *Presse méd 2* 1915–1918 1936
129 LABORDE, S Les dangers des substances radio-actives introduites dans l or ganisme. *Presse méd 47*.393–394 1939
130 LACAPÈRE, G : Etude sur l'ostéophytose vertébrale. *Rev rhum 14*.249 259 1947 and *15* 17–20 1948

131 LAGAPÈRE, J Etude sur l ostéophytose vertébrale. *Acta physiother rheum. belg* *4* 145–159 1949
132. LAGASSAGNE, A. (Quoted by Coste, F [See Ref. 48])
133 LA CHAPELLE, E. H Osteotomy of the lumbar spine for the correction of kyphosis in cases of ankylosing spondylarthritis. *J Bone & Joint Surg* 28 851–858 1946
134 LAW W A. Post-operative study of vitallium mould arthroplasty of the hip joint. *J Bone & Joint Surg.*, *30-B* 76–83 1948
135. LÉRI A. La spondylose rhizomélique (étude d'ensemble). *Rev méd.*, *19*.597–624 691–733, 801–833 1899
136 LÉRI, A. Contribution à l étude de la pathologie du rachis. Autopsie d un cas de cyphose hérédo-traumatique. *Bull et mém Soc méd hôp Paris 31*.875–880 1904
137 LÉRI, A. *Études Sur les Affections de la Colonne Vertébrale* Paris, Masson 1926
138 LÉRI, A. and LIÈVRE, A. A. L action de la médication radioactive (thorium X) sur le psoriasis et les arthropathies psoriasiques. *Bull et mém. Soc m d hôp Paris 53* 1335–1339 1929
139 LÉRI A. and THOMAS Le thorium X dans le traitement des rhumatismes chroniques. *Bull. méd Paris*, *36*.348–356 1922
140 LIÈVRE, J A. Bilan des opérations parathyroïdiennes dans le rhumatisme chronique. *Rev rhum.*, *16*·447–466, 1949
141 LUDWIG A. O, SHORT C. L. and BAUER W Rheumatoid arthritis as a cause of increased cerebrospinal fluid protein study of 101 patients. *New England J Med* 222.306–310, 1943
142. MARIE, P and ASTIÉ, C. Sur un cas de cyphose hérédotraumatique. *Presse méd.* *5*.205–206, 1897
143. MARIE, P Sur la spondylose rhizomélique. *Rev méd* *18*.285–315 1898
144 MARIE, P and LÉRI, A. La spondylose rhizomélique. Anatomie pathologique et pathogénie. *Nouv Iconogr Salpêtrière 19*.32–49 1906
145 MATHIEU P and LANGE, P La sacro-coxalogie chez l'adulte. *Rev méd. franç.*, *15*.257–262, 1934
146 MENNELL, J *Backache* London Churchill, 1931
7 MEYER, M. and FORSTER, E. Considérations pathogéniques sur "l'hyperostose monilliforme du flanc droit de la colonne dorsale. *Rev rhum. 5*.286–294 1938
8 OPPEL, V A. Parathyroidectomy for ankylosing polyarthritis. *Ann. Surg 90* 978–981 1929
9 OPPENHEIMER, ALBERT Diseases of the apophyseal (intervertebral) articulations. *J Bone & Joint Surg* *20*.285–313 1938
0 OPPENHEIMER, ALBERT Calcification and ossification of the vertebral ligaments (spondylitis ossificans ligamentosa) roentgen study of pathogenesis, and clinical significance. *Radiology 38* 160–173 1942.
151 OPPENHEIMER, ALBERT Paravertebral Abscesses associated with Strümpell Marie disease. *J Bone & Joint Surg 25 N S*·90–96 1943
152. OPPENHEIMER, Albert Development, clinical manifestations and treatment of rheumatoid arthritis of the apophyseal intervertebral joints. *Am. J Roentgenol.*, *49* 49–76, 1943
153 OVERGAARD K. On Bechterew's disease from the roentgenologic point of view *Acta radiol.*, *26* 185–209 1945
154 PARR, L. J A. and SHIPTON E. Spondylitis ankylopoetica. *M J Australia*, *1*.277–290 1946.

155 PERMANYER, J J and BARCELO P Modificaciones hematicas en la espondiloartritis anquilopoyetica examen morfologico y cuadro proteico. *Rev españ. reum.*, *1* 238–244 1945

156. PIÑOL AGUADE, J., BARCELÓ P and ROTES-QUEROL, J Polyartritis cronica y psoriasis. *Rev españ. reum.* 2 373–403 1948.

157 PLANTIVEAU P Intoxication par le fluor (Thesis No. 261) Paris, 1946 110 p.

158 POLLEY H. F and SLOCUMB, C. H Rheumatoid spondylitis a study of 1035 cases. *Ann. Int. Med.* *26* 240–249 1946

159 POIRIER, P and CHARPY A. Sur le ligament vertébral antérieur *Traité d'anatomie humaine* 3rd. Ed. *1* Paris, Masson, 1911

160 POSTH, MAC Le sacrum description, développement anomalies, articulations (Thesis). Paris, 1897

161 RACE (Quoted by Buckley C. W [See Ref. 32]).

162. RAGAN C. and BOOTS, R. H Treatment of gold dermatitides use of B.A.L. (2,3-dimercaptopropanol). *J.A.M.A.*, *133* 752–754 1947

163. RAINER, F S. *L œuvre Scientifique de F S. Rainer II Structure Fonctionnelle.* Bucarest, Imprimerie Nationale 1945

164 RAMBAUD A. and RENAULT C. *Origine et Développement des Os* Paris, Chamerot, 1864

165. RAY M. Spondylitis in twins. *Proc Roy Soc Med.*, *25* 325–326, 1932.

166. RENDICH, R. A. and Shapiro, A. V Osteitis condensans ilii. *J Bone & Joint Surg* *18*:899–908 1936

167 QUERY R. Z., JR. Rheumatoid spondylitis in early diagnostic features and management. *J.A.M.A.* *139* 692–698 1949

168 Riebel, F Ossifications of the costal cartilages their relation to habitus and disease. *Am. J Roentgenol* *21* 44–47 1929

169 RIST E. GALLY L. and TROCMÉ, C. L ossification des cartilages costaux dans l espèce humaine étude d'anatomo-radiologie statistique. *Presse méd.*, *36* 641–644 1928

170 ROBINSON, W D Personal communication to Smyth, C. J In Comroe, B. I *Arthritis and Allied Conditions* 4th Ed. Philadelphia, Lea & Febiger 1949 107

171 ROGEN A. S The heart in rheumatoid arthritis. *Brit. M J.*, *1* 87–88 1947

172. ROGERS M H. and CLEAVES, E. N Adolescent sacroiliac joint syndrome. *J Bone & Joint Surg.*, *17* 759–768, 1935

173 ROGOFF B and FREYBERG R. H. The familial incidence of rheumatoid spondylitis. *Ann. Rheumat Dis.*, *8* 139–142, 1949

174 ROHOLM, K. *Fluorine Intoxication, A Clinical Hygienic Study With a Review of the Literature and Some Experimental Investigations* Copenhagen, Lewis, 1937

175 ROPES M W ROBERTSON W V B., ROSSMEISL, E. C., PEABODY R. B. and BAUER W Synovial fluid mucin. *Acta med. scandinav Suppl* *196* 700–744 1947

176 ROQUES E. and ARLET J Les manifestations articulaires périphériques de la spondylarthrite ankylosante. *Rhumatologie* No. 4 168, 1949

177 ROSENBERG E. F BAGGENSTOSS, A. H and HENCH, P S Causes of death in 30 cases of rheumatoid arthritis. *Ann. Int. Med* *20* 903–919 1944

178 ROSENBERG E. F HENCH, P S BISHOP L. F and WEINTRAUB H J Cardiac lesions in rheumatoid arthritis. A summary of recent developments and a bed-side study of patients and controls. *Arch. Int Med* *85* 751–764 1950

179 ROTES-QUEROL, J Espondilartritis anquilopoietica y tuberculosis. *Rev españ. reum* *1* 226–273 1945.

180 ROTES-QUEROL, J Les calcifications précoces des cartilages costaux dans la spondylarthrite ankylosante *Rev rhum* 17 314–315 1950
181 ROTES-QUEROL, J Les débuts de la spondylarthrite ankylosante *Toulouse méd* 15–32, 1950
182 ROUD A. *Mécanisme Des Articulations et Des Muscles de l'Homme* Paris Baillère 1913
183 ROUVIÈRE, H *Anatomie Humaine Descriptive et Topographique* Paris Masson, 1943
184 RUFFER, A. Studies in palaeopathology arthritis deformans and spondylitis in ancient Egypt. *J Path. & Bact.* 22 152–196 1918–1919
185 SAIDMAN J *Diagnostic et Traitement Des Maladies de la Colonne Vertébrale* Paris Doin, 1948
186 SASHIN D A critical analysis of the anatomy and the pathologic changes of the sacro-iliac joints. *J Bone & Joint Surg* 12:891–910 1930
187 SCHEUERMANN H Roentgenologic studies of the origin and development of juvenile kyphosis, together with some investigations concerning vertebral epiphysis in man and in animals. *Acta orthop scandinav* 5 161–216 1934
188 SCHINZ, H. R. BAENSCH W and FRIEDL, E. *Lehrbuch der Röntgendiagnostik*, 4th Ed. Leipzig Thieme 1939
189 SCHMORL, G *Archiv und Atlas der normalen und pathologischen Anatomie in typischen Röntgenbildern. Die Gesunde und Kranke Wirbelsäule im Röntgenbild* Leipzig Thieme 1932.
190 SCOTT S G X ray in chronic rheumatic arthritis diagnosis, prognosis and treatment. *Brit. J Phys Med* 10 127–128 1935
191 SCOTT S. G Chronic infection of the sacro-iliac joints as a possible cause of spondylitis adolescens. *Brit J Radiol* 9 126–131 1936
192. SCOTT S. G *A Monograph On Adolescent Spondylitis or Ankylosing Spondylitis The Early Diagnosis and Its Treatment By Widefield X-ray Irradiation.* Published under the auspices of the Nuffield X-ray Therapy Research No. 1 London, Oxford University Press 1942.
193 DE SÈZE, S *et al* *Algies Vertébrales d'Origine Statique (Région Lombaire et Lombo-sacrée).* Paris, Expansion Scientifique Française, 1949
194 SÈZE, S. DE, ROTES-QUEROL, J and DJIAN A. Le diagnostic radiologique de la hernie discale postérieure en station verticale. Valeur diagnostique du baillement discal électif provoqué. *Semaine hôp Paris* 26 1297–1307 1950.
195 SICARD J A. GALLY and HAGUENEAU Ostéites condensantes à étiologie inconnue. *J Radiol. et électrol* 10 503–507 1926
196 SIMON R. Les traitements chirurgicaux de la spondylose rhizomélique. *Rev rhum.*, 2 409–451 1935
197 SIMPSON N R. W and STEVENSON C. J An analysis of 200 cases of ankylosing spondylitis. *Brit M J* 1 214–216 1949
198 SIMPSON N R. W KERSLEY G D and BROOKS D H. Effect of blood transfusion on rheumatoid arthritis. *Ann. Rheumat Dis* 8 277–284 1949
199 SIVÉN V O Zur Kenntnis der sogen. chronisch ankylosirenden Entzündung der Wirbelsäule. *Ztschr klin. Med.*, 49 341–376 1903.
200 SMITH PETERSEN M. N Clinical diagnosis of common sacroiliac conditions. *Am. J Roentgenol* 12 546–550 1924
201 SMITH-PETERSEN M. N Arthroplasty of the hip. A new method. *J Bone & Joint Surg* 21 269–288, 1939

202. Smith Petersen M. N Larson C. B. and Aufranc, O E. Osteotomy of the spine for correction of flexion deformity in rheumatoid arthritis. *J Bone & Joint Surg* 27 1–11 1945
203 Smith-Petersen M. N Evolution of mould arthroplasty of the hip joint. *J Bone & Joint Surg* 30-B 59–75 1948
204 Smyth, C. J Freyberg R. H. and Peck, W S Roentgentherapy for rheumatic diseases. *J.A.M.A.* 116 1995–2001 1941
205 Smyth, C. J Freyberg R H. and Lampe, I. Roentgentherapy for rheumatoid arthritis of the spine (Marie-Strümpell arthritis spondylite rhizomélique). *J.A.M.A.*, 117 826–831 1941
206 Stecher, R. M., Solomon W M. and Wolpaw R. Heredity in rheumatoid and ankylosing spondylitis. *Rheumatic diseases based on the proceedings of the Seventh International Congress on Rheumatic diseases, 1949* Philadelphia, Saunders, 1952, 66–67
207 Steiner, G., Freund, H. A. Leichtentritt B. and Mann M. E. Lesions of the skeletal muscles in rheumatoid arthritis nodular polymyositis. *Am. J Path.* 22 103–145 1946
208 Strümpell, A. *Lehrbuch der Speciellen Pathologie und Therapie der Inneren Krankheiten.* Leipzig, Vogel, 1884
209 Strümpell, A. Bemerkung über die chronische ankylosierende Entzündung der Wirbelsaüle und der Hüftgelenken. *Deutsche Ztschr Nervenh.* 11 338 1897
210 Swaim L. T and Kuhns J G Prevention of deformities in chronic arthritis, spine and head. *J.A M.A.* 94 1123–1125 1930
211 Swaim, L. T Le traitement orthopédique du rhumatisme chronique. *Conférence scientifique internationale du rhumatisme chronique progressif généralisé.* Aix les-Bains, 1934 Chambéry Imprimeries Réunies, 1934 505–533
212. Swaim L. T Orthopaedic treatment of Strümpell-Marie arthritis. *J Bone & Joint Surg.*, 21 983–991 1939
213 Vilaseca Sabater, J M. and Barcelo P *Patologia de las Pequenas Articulaciones Intervertebrales* Barcelona, Salvat, 1946.
214 Vuillermet *La panarthrite engainante* (Thesis). Lyon, 1940
215 Weil, M. P and Oumansky V Qu est-ce que la spondylose rhizomélique? *Rev méd.*, 52 327–345, 1935
216 Weil, M. P and Colliez, R. La radiographie de l articulation sacro-iliaque. *Cahiers radiol* 3 241–249 1937
217 Weil, M. P and Sichère, R. M. La spondylite rhumatismale à évolution lente et à révélation tardive. *Rev rhum.* 16 263–270 1949
218 West H. F The aetiology of ankylosing spondylitis. *Ann. Rheumat. Dis.*, 8 143–148 1949
219 Williams, A. J Rheumatoid (Marie-Strümpell) spondylitis. Technique of examination and importance of the costal joints. *California Med.*, 70 257–261 1949
220 Wyatt H. Ankylosing spondylitis. *Brit J Radiol* 18 301–302, 1945
221 Young J Pelvic osteo-arthropathy in pregnancy *Proc Roy Soc Med.*, 32 1591–1597 1939

ADDITIONAL BIBLIOGRAPHY (Compiled by the Authors since 1950)

222. Amoudruz, J P Etude anatomique et pathogénique de l'ostéophytose des corps vertébraux. Thesis, Paris, 1951

223 BALL, J Sheep cell agglutination for rheumatoid arthritis. *Ann Rheumat Dis* *11*:97–111 1952.

224 GEIRMANN E. W Fractures of the ankylosed spine. *J Bone & Joint Surg* *31* 4:669–671 1949

225 BOOTS R. H LIPMAN M O COSS, J A. AND RAGAN CH Immunologic re actions in rheumatoid arthritis. *Rheumatic Diseases* Philadelphia, Saunders 1952, pp. 336–341

226. CECIL, R. L., NICHOLLS E. E. AND STAINSBY W J Etiology of rheumatoid arthritis. *Am. J M Sc* *181* 12–25 1931

227 COSTE, F CAYLA, J AND DELBARRE, F Cortisone et corticostimuline (ACTH) en rhumatologie. Paris, Masson 1953

228 CRUICKSHANK, B Histopathology of diarthrodial joints in ankylosing spondylitis. *Ann. Rheumat. Dis.*, *10*.393–404 1951

229 *Ibid* Interpretation of multiple biopsies of synovial tissue in rheumatic diseases. *Ann Rheumat. Dis* *11* 137–145 1952.

230 DAVIS, J B. AND BLAIR H C. Spurs of the calcaneus in Strümpell Marie disease. *J Bone & Joint Surg.*, *32 A*.834–840 1950

231 DAVISON R. A. Relationship of clinical findings to urinary ketosteroid in ankylosing spondylarthritis and rheumatoid arthritis. Il Congreso europea de reumatologia, Barcelona, 1951 *Communicaciones*, pp. 7–10

232. EFFERSOE, P The serum proteins in rheumatic diseases. Il Congreso europea de reumatologia, Barcelona, 1951 *Communicaciones*, pp. 504–508

233 FORESTIER, J JACQUELINE, F AND ROTES-QUEROL, J Spondylarthrite ankylosante à évolution rachidienne indolore. *Rev rhum.* *18* 486–487 1951

234 FORESTIER, J CERTONCINY A. JACQUELINE, F and ROTES-QUEROL, J La spondylarthrite ankylosante. Il Congreso europea de reumatologia, Barcelona, 1951 *Ponencias oficiales* pp 3–58

235 FORESTIER, J JACQUELINE, F and BEWS A. Exploration du couple hypophyse-surrénal chez les sujets atteints de spondylarthrite ankylosante. *Presse méd.*, *60*.298–299 1952.

236 FORESTIER, J JACQUELINE, F and CANET L. Aspects radiologiques des poignets et des mains dans les polyarthrites chroniques de l adulte et la spondylarthrite ankylosante. *J radiol et Electrol* *33*.341–348 1952.

237 FORESTIER, J and JACQUELINE, F La polyarthrite masculine. *Ann. Rheumat. Dis* *11*.264–271 1952.

238 FRANÇON F JOLY L., LAC, DU G and TOUSSAINT J Les lésions osseuses de la symphyse pubienne dans la spondylarthrite ankylosante. *Presse méd.* *60* 436–438 1952.

239 FRIEDENWALD J S Ophthalmic Pathology Philadelphia, Saunders 1952, p 126.

240 HADLEY L. A. Accessory sacro-iliac articulations. *J Bone & Joint Surg* *34-A* 149–155 1952.

241 HART F D BOGDANOVITCH A. and NICHOL, W D The thorax in ankylosing spondylitis. *Ann Rheumat. Dis.*, *9* 116–131 1950

242. HERBERT J J Techniques et résultats des ostéotomies vertébrales d après 26 observations. *Acta Orthop Scandinav.*, *22*.36–58 1952.

243 HOLLANDER, J L. BROWN E. M. JR., JESSAR, R. A. and BROWN C. Y Hydrocortisone and cortisone injected into arthritic joints. Comparative effects of and use of hydro-cortisone as a local anti-arthritic agent. *J.A.M.A.* *147* 1629–1635 1951

244 JACQUELINE, F., DEPRAITÈRE, R. and TRUET J Modifications des protides dans les rhumatismes chroniques inflammatoires. *Rev rhum.* 18.527–532, 1951

245 JACQUELINE, F and ROTES-QUEROL, J Ponts osseux juxta-discaux. Etude macroscopique d'une pièce de spondylarthrite ankylosante. *Rev rhum.* 19 446–452, 1952.

246 JACQUELINE, F Etude comparative des altérations radiologiques des hanches dans la polyarthrite chronique évolutive de l'adulte et la spondylarthrite ankylosante. *J radiol. et Electrol.* 33.219–222, 1952.

247 JACQUELINE, F EYQUEM, A. and REBEYROTTE, P Recherches sérologiques dans les rhumatismes chroniques inflammatoires. *Rev rhum.* 19·928–932, 1952.

248 KALBAK, K. Agglutination of hemolytic streptococci (group A) in serum from patients with rheumatoid arthritis. *Ann. Rheumat. Dis* 6.230–240 1947

249 KNUTSON F Changes in the sacro-iliac joints in morbus Bechterew and osteitis condensans ilii. *Acta Radiol* 33 557–569 1950

250 KOCH, W Der Nachweis von Thorium X in der Wachstumsfüge und sein Einfluss auf das Langenwachtum beim jügendlichen Kaninchen. *Orthopädie Grenzgeb* 80.532–546, 1951

251 *Ibid* Die Verteilung von Peteosthor und seiner Hauptbestandteile Thorium X und Platin im heranwachsenden Organismus und der Einfluss von Thorium X auf das Fügenwachstum beim jügenlichen Kaninchen. *Strahlentherapie* 85.253–289 1951

252. LOGHEM LANGEREIS, P E. VAN Serologische Reacties bij rheumatische Ziekten. Thesis Amsterdam, 950

253 OTT V R. De l'hyperostose ankylosante vertébrale sénile. *Rheumatol* No. 3 pp 104–109 1952.

254 PONS, A. BARCELO P VILASECA-SABATER, J M. ROTES-QUEROL, J and BATTALLA BOIXET J E. Manifestationes osteoarticulares de la brucellosis. II Congreso europea de reumatologia, Barcelona, 1951 *Ponencias officiales* pp. 395–503

255 QUINN R. W and LIAO S J Studies on the differential diagnosis of adult rheumatic fever and rheumatoid arthiritis by means of serologic tests. *Rheumatic Diseases* Philadelphia, Saunders 1952, pp. 331–335

256 ROKITANSKY C. *Lehrbuch der Pathologische Anatomie* Vienna, Braûmuller 1956, 3rd ed. (quoted by Ott, p. 253).

257 ROMANUS R and YDEN, S Destructive and ossifying spondylitic changes in rheumatoid ankylosing spondylitis. *Acta orthop scandinav* 22:88–99 1952.

258 RUTISHAUSER, E. FORESTIER J HERBERT J J RABINOWICZ, TH. and GRASSET E. Aspects anatomo-cliniques des arthropathies de la hanche. *Rev rhum.* 19.869–898 1952

259 SAVILL, D L. The manubrio-sternal joint in ankylosing spondylitis. *J Bone & Joint Surg* 33-B 56–64 1951

260 SCHELTER, G Alterations of the serum proteins in perispondylarthritis ankylopoietica. II Congreso europea de reumatologia. Barcelona, 1951 *Communicaciones*, pp 41–43

261 SHERMAN M S Psoriatic arthritis. *J Bone & Joint Surg.*, 34-A.830–852, 1952.

262. SHIPP F L. and HAGGART G E. Further experience in the management of osteitis condensans ilii *J Bone & Joint Surg* 32 A:841–847 1950.

263 SOHOLT S T Tuberculous of the sacro-iliac a review of seventy-five cases. *J Bone & Joint Surg.*, 33-A 119–130 1951

264 STECHER, R. M. and HAUSER, H. Ankylosing spondylitis in two pairs of sisters with comments on heredity and pregnancy. II Congreso europea de reumatologia. Barcelona, 1951. *Communicaciones* pp. 3–7

265 SWAAY VAN H. Spondylosis ankylopoietica en pathogenetische studie. Thesis. Leyden, 1950

266 *Ibid.* The pathology of ankylosing spondylitis. II Congreso europea de reumatologia. Barcelona, 1951. *Ponencias officiales* pp. 99–115

267 WAALER O. On the factor producing agglutination of sensitized red cells and its relation to the agglutination of hemolytic streptococci in rheumatoid arthritis sera. *Ann. med. exper. et biol. Fenniae* 28 Suppl. 8 1950

268. WENZEL, C. *Uber die Krankheiten am Rückgrate.* Bamberg 1824 (quoted by Schmorl and Junghanns [189]).

269 WETTSTEIN P. and RIOTTON G. Aspects anatomo-radiologiques de la spondylarthrite ankylosante. *Radiol. Clin.* 19 325–331 1950

270 WOODS A. C. Clinical and experimental observation on the use of ACTH and Cortisone in ocular inflammatory diseases. *Am. J. Ophth.* 33 1325–1349 1950.

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This Book

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By J FORESTIER, F JACQUELINE *and* J ROTESQUEROL

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